

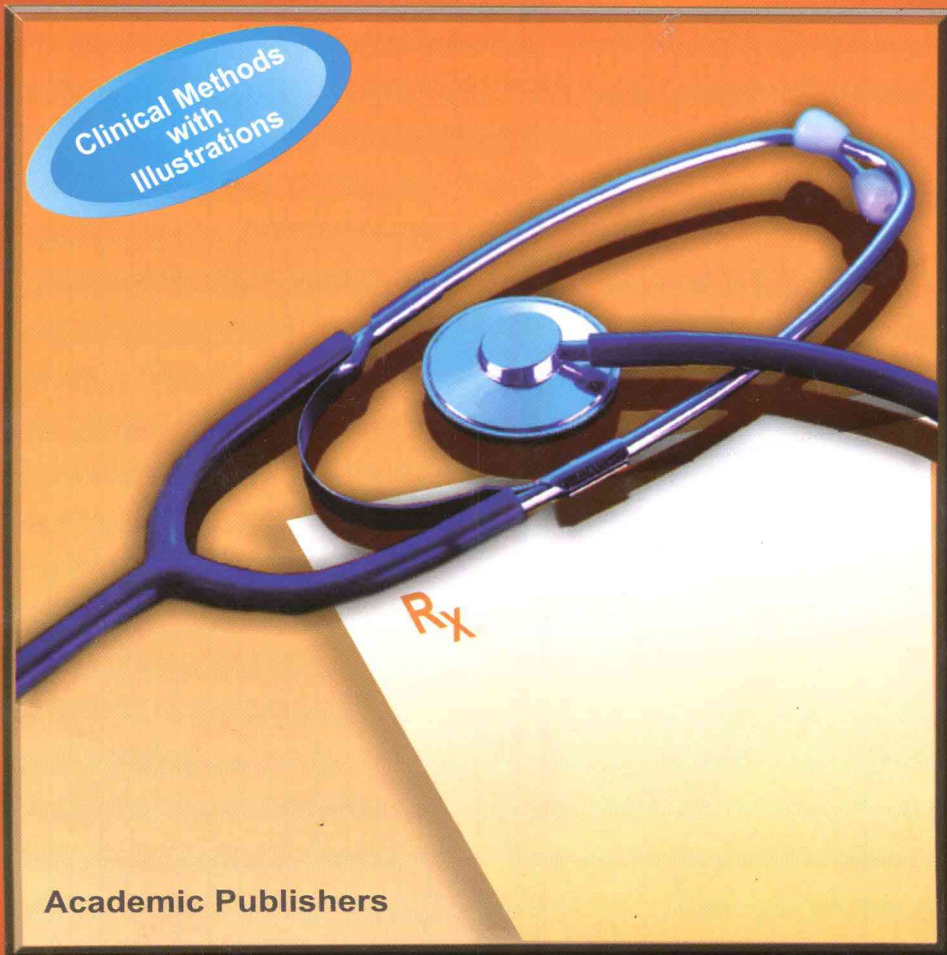
6th Edition

Bedside Clinics in Medicine

Part I

Long Short Spot Cases

Arup Kumar Kundu



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Author : Fellow/Member of

- Indian College of Physicians (Ind)
- New York Academy of Sciences (USA)
- International Advisory Panel of "Kumar & Clark's"
Textbook, 'Clinical Medicine', 7th Edition (Edinburgh)
- Association of Physicians of India (API)
- Indian Rheumatology Association (IRA)
- Indian Medical Association (IMA)

“Medicine is to be learned only by experience; it is not an inheritance; it cannot be revealed. Learn to see, learn to hear, learn to feel, learn to smell, and know that by practice alone can you become expert. Medicine is learned by the bedside and not in the classroom. See, and then reason and compare and control. But see first.”

— **Sir William Osier (1849-1919)**

Professor of Medicine, Oxford, UK

BEDSIDE CLINICS IN MEDICINE

PART I

SIXTH EDITION

Long Short & Spot Cases in Medicine
(Includes 37 Tables & 193 Coloured Photographs)

Arup Kumar Kundu md frcp mnas

Professor, Department of Medicine

KPC Medical College, Kolkata, India

Formerly Professor of Medicine & In-charge, Division of Rheumatology

R. G. Kar Medical College, Kolkata, India

Formerly Assoc. Prof. of Medicine & In-charge, Division of Rheumatology

N. R. S Medical College, Kolkata, India

Author of:

Bedside Clinics in Medicine, Part II

MCQs in Internal Medicine

Pearls in Medicine for Students

Chapter in API Textbook of Medicine, 8th Edition

Chapter in Postgraduate Medicine, 2009

Chapters in 'Rheumatology : Principles and Practice', 2010

Chapters in Medicine Update : 2010 & 2011

&

Section on Online Appendix of "Kumar & Clark's"

Textbook, 'Clinical Medicine', 6th & 7th Edition



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BEDSIDE CLINICS IN MEDICINE, PART I

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Price : Rs. 460.00

Dedicated to my father

Professor (Dr.) AMIYA KUMAR KUNDU, MD (Cal)

a great clinician

Who inspired me to learn clinical medicine

A few personal communications with Giants of Medicine

(i)

From :

Dr. John Macleod
13 Merchiston Avenue
Edinburgh, Scotland, EH 10 4PJ

(Regarding an error in renal clearance test)

13/7/83

Dear Dr. Kundu,

It was good of you to take the trouble to write to me about the 13th edition of Davidson.

Thank you for your kind comments.

You are of course, correct about the typographical error on page 428. It should have been $C = U \times V / P$; the printer omitted the division sign. This is now being adjusted.

Again many thanks for writing.

With best wishes.

Yours sincerely

Sd/ **John Macleod**

(2)

From :

Kurt J. Isselbacher, M.D.
Director, Massachusetts General Hospital Cancer Center, Boston
Mallinckrodt Professor of Medicine, Harvard Medical School
Editor : Harrison's Principles of Internal Medicine. 13th Edition

(Regarding errors in serum-ascites albumin gradient)

February 23, 1998

Dear Dr. Kundu,

I must commend you for your astute and thorough reading of the Harrison's textbook and in addition, thank you for pointing out two obvious typographical errors. In fact, this was already done in the second printing of the 14th edition.....

Again, many thanks for your thoughtfulness in bringing these two typographical errors to our attention.

Sincerely,

Sd/ **Kurt J. Isselbacher**

(3)

From :

Keith A A Fox
Duke of Edinburgh, Professor of Cardiology and Head of Department
Cardiovascular Research Unit. The University of Edinburgh
Hugh Robson Building. George Square, Edinburgh EH8 9XF

(Regarding erythema nodosum, & pulmonary stenosis)

5th December 1995

Dear Dr. Kundu,

Re : Davidson's Principles and Practice of Medicine : 1 7th Edition 1995

On behalf of Editors of this textbook I have been asked to respond to you in view of the fact that this chapter was written by Dr. Boon and myself.

In response to your first question we have obtained source material which indicates erythema nodosum is a manifestation of acute rheumatic fever.

In conjunction with pulmonary stenosis, the only reason why the thrill may be best palpated in expiration is because of the fact that the heart moves closer to the anterior chest wall. We entirely agree that the velocity of blood flow across the pulmonary valve is increased in inspiration and this is demonstrated using echo Doppler techniques. However, for the reasons mentioned above palpation of thrill may be more obvious in held expiration when the heart and vessels are closer to the chest wall. I hope that these comments help to clarify the situation.

Yours sincerely

Sd/ **Keith A A Fox**

Professor of Cardiology

(4)

From:

Ellen Green
Senior Commissioning Editor
Elsevier. 1 -3 Baxter's Place
Leith Walk. Edinburgh EH1 3AF

(Regarding contribution in "Kumar & Clark's" textbook on Medicine)

23/6/2005

Dear Dr. Kundu

Firstly, on behalf of Kumar & Clark, I would like to thank you for your contributions to the Online Appendix for **Clinical Medicine 6e** I hope you are pleased with the book Kind regards.

Sd/ **Ellen Green**

(5)

From:

(Regarding some clinical methods & apex beat localisation)

Dr. J G Douglas
Chest Clinic, Clinic C
Aberdeen Royal Infirmary, Foresterhill
Aberdeen AB25 2ZN

2nd November 2005

Dear Dr. Kundu

Ref: MacLeod's Clinical Examination. 11th Edition

Thank you very much for your recent e-mail I am an editor of this edition..... I am responsible for writing much of the Respiratory Chapter and was most interested in your comments. In response:

Examining for Tracheal Shift..... Clearly this technique could also be performed with the patient standing or sitting.

Technique of Percussion - I entirely agree with your comment that to obtain the loudest percussion note ideally the other fingers, apart from the middle finger, should not touch the skin surface. Figure 4.19 is ambiguous and I will make a note of this for the next (12th) edition.

... Vocal Resonance..... However I agree that as written in the first paragraph of page 143 there is some confusion..... Again I will make a note to clarify this for the next (12th) edition.

Aegophony (page 144) However I agree that it's position is a little confusing and again I will make a note of this when redrafting the next (12th) edition.

Examination Sequence (page 97 palpation of apex beat) - I entirely agree with you that the second half of the statement under the 3rd arrow: "If you cannot feel it, ask the patient to roll on to their left side" is incorrect and references the wrong figure! I will make sure this is removed at the next reprint.

We are enormously grateful to you for your attention to detail in pointing out these points.

With best wishes. Yours sincerely,

Sd/ J G Douglas

(6)

From :

(A compliment from Chief Editor of Harrison's book)

Dennis L. Kasper. M.D.
William Ellery Channing Professor of Medicine and
Professor of microbiology and Molecular genetics
Harvard Medical School
Director. Channing Laboratory
Brigham and Women's Hospital. Boston. MA 02115
Editor : Harrison's Principles of Internal Medicine. 16th edition.

7/10/2006

Dear Dr. Kundu,

..... I very much appreciate your interest in contributing to Harrison's. I have looked up your publications on PubMed, and I can see that you are active and productive in your field..... and I wish you all the best in your ongoing studies and clinical practice.

Best regards,

Sd/ Dennis Kasper

(7)

From :

(Regarding some queries on clinical medicine)

Laurence Hunter.
Senior Commissioning Editor. Elsevier

11/9/2008

Dear Dr. Kundu,

ReJ: Hutchison's Clinical Methods. 22 Ed

Once again I am most grateful to you for taking the trouble to contact us with queries about the content of this textbook. I shall pass on to the Editors the comments and ask them to take full account of these in the revision of the book.

Thank you once again for your interest in our publishing programme.

With all good wishes.

Sd/ Laurence Hunter

Foreword

It is a pleasant task for me to introduce Dr. Arup Kumar Kundu, MD (Cal) who is known to me as a brilliant student since his undergraduate days.

He had his postgraduation under my direct supervision and I found him extremely hardworking and very sincere. His clinical acumen was superb. Dr. Kundu was one of my best students I had in my teaching career. I had every reason to believe that Dr. Kundu would be a very good teacher in Clinical Medicine in future. Today my expectation has come true.

With the advent of newer and more sophisticated investigative procedures, importance of Clinical Medicine seems to be on the wane at present. But in our country, costly investigations are out of reach to most of the patients. Naturally, Clinical Medicine as basic diagnostic procedure will continue to be important in our country for a long time.

While conducting Oral & Practical part of MBBS examination, Dr. Kundu has attained sufficient knowledge about its protocol. With the help of this knowledge he has written the book—"Bedside Clinics in Medicine" to guide students. He has made a comprehensive list of patients that are given as 'cases' in the examination and has discussed meticulously the different questions a student might encounter with their satisfactory answers.

I am sure, this book will be of great help to both undergraduate and postgraduate students. I wish him all success.

Date : 12.10.96
Kolkata

DR. SUBRATA SEN
MBBS (Cal), FICA (USA), FRCP (Edin),
Ex-Professor Director, Department of Medicine,
Institute of Postgraduate Medical Education
and Research (SSKM Hospital, Calcutta)

Preface to the First Edition

This is not a text book of medicine. This book is based on bedside clinics on hundred 'model cases' oriented to **oral and practical examinations** with special stress on clinical methods. I hope this book will be very much helpful to students of clinical medicine both undergraduate and postgraduate.

First, I wish to express my deep sense of gratitude to my parents and family members, whose constant encouragement has enabled me to complete this book.

Next, I take the opportunity to extend my heartfelt reverence and fathomless gratitude to all my respected teachers. I feel, I should make a special mention of the names of Prof. P. Sen, MD (Cal), FRCP (Edin), FICA (USA), Prof. S. Sen, FRCP (Edin), FICA (USA), Prof. P. C. Das, MD (Cal), FICA (USA), FCCP (USA), and Prof. A. K. Kundu, MD (Cal).

This book would never have been completed without the help of my wife Mrs. Bijoya Kundu. I am greatly indebted to her.

I owe deep sense of gratitude to Prof. S. K. Paul, MO (Cal) without whose constant encouragement, active guidance and blessings, this book would never have seen the light of the day.

I am very much grateful to Mr. Bimal Dhur of Academic Publishers, whose advice has been of great help to me.

I am thankful to Mr. Robin Chatterjee and Ms. Lopamudra Paul for meticulously typing out the manuscript. I also heartily acknowledge Mr. Kaustuv Paul of Crest Publishers who has taken immense pain in publishing this book.

I also express my thanks to all the staff of 'Abhinaba Mudrani' who have helped me in every step to complete the book.

In conclusion, I would like to admit that only few names appear in this acknowledgement, many too much unsung, have significantly contributed to and enhanced the quality of this book. To all, I am deeply obligated.

Date : 12th October, 1996

"Trimurti"

BG-87, Sector-II, Salt Lake City,
Kolkata-700 091, India.

Arup Kumar Kundu

Preface to the Sixth Edition

It gives me immense pleasure to present the sixth edition of the book in a new get-up. The overwhelming success of the previous editions has encouraged me to bring out the present thoroughly revised new edition. Most of the chapters have been rewritten and significantly revised maintaining the style and old tradition intact. The book reviews a wide range of common clinical cases with discussion on core topics. Many new photographs have enriched the 'colour atlas' section while replacing majority of older ones. I expect that the 'clinical methods atlas' section will prove to be immensely helpful for aspiring students.

I do believe, bedside medicine still holds strong in the era of high-tech medicine and this is why I have attempted to write this book dealing with common clinical problems as well as some fundamental information which the students are expected to know at the end of their clinical training. Even with the availability of modern gadgets to have advanced and accurate investigations, clinical examination is very important in reaching a logical diagnosis. I have retained the original objectives of the book — totally clinically oriented, concise but comprehensive, and as far as practical with a lucid language. The new edition deals with different must-know areas in clinical medicine in a student-friendly way keeping the methodical and logical approach unaltered. The problem-solving attitude will help the students in their oral and practical, as well as theoretical examination, and also in their professional life in future. I think that this monograph is a quick-reference ready-reckoner handbook and a learning-revision tool to increase the core knowledge.

Other than the new look and new photographs, the new edition also includes two very basic chapters on 'cyanotic congenital heart diseases' and 'polyarthrititis'. The book remains an invaluable resource for undergraduates, house officers preparing for MD/DNB/MRCP, and the postgraduates, and I hope that the junior teachers will also be benefited by reaching a high level of clinical competence if they go through it. As clinical methods are scattered throughout the book, the students are advised to follow the 'index' for easy reading. I feel happy and jubilant as this book was nicely reviewed by renowned indexed journals, leading physicians, students and many academicians from different corners of the country. However, healthy suggestions and constructive criticisms through e-mail (arup.kundu@hotmail.com) will always be appreciated by the author.

I would like to record my appreciation for Mr. Kaustuv Paul of KSP Udyog for publishing, Mr. Bimal Dhur and Dipankar Dhur of Academic Publishers for distributing, and Mr. Amar Nandy for printing the book with great care. I am grateful to all my patients whose photographs have been printed in this book and to the MSVP, R. G. Kar Medical College & Hospital, Kolkata for permitting me to take the photographs. This work would not have been possible without the constant support and encouragement from my family members, specially to speak of my wife Bijoya Kundu, my daughter Ushasi and my son Abhishek, which ultimately made this edition a reality.

Date : 15th April, 2010

"Trimurti"

BG-87. Sector-II, Salt Lake City,
Kolkata-700 091, India.

e-mail : arup.kundu@hotmail.com

Arup Kumar Kundu

In their esteemed opinion about the book

• “Your book is an excellent replicative educational medium for exam-going students. Congratulations

• “..... I am proud of you as you are a THINKER. May your tribe increase. Many students make you their role model. God will bless you always.....” —Prof. B. M. Hegde, Ex-Professor & Dean of Medicine, Kasturba Medical College, Mangalore (Ex-Vice-Chancellor, MAHE University, Manipal).

“... Dr. Kundu has brought out a book based on bedside clinics on 26 model long cases and 74 short and spot cases oriented to clinical and oral examinations with special stress on clinical methods..... He discusses the history, diagnosis, signs, differential diagnosis and management in the form of questions and answers. The answers are given in detail. There is an exhaustive coverage of the subject and Dr. Kundu has to be congratulated for bringing out such a wealth of knowledge. This is not a text book but it contains a lot of information which the students are expected to know at the end of the clinical training..... The book is very helpful to the students of clinical medicine while revising the subject before examination. Dr. Kundu has discussed the various questions which student may encounter during the examination and he has done it admirably”. —Book review in Journal of the Association of Physicians of India (JAPI) by Dr. P. S. Shankar, Dean, K. J. Somiya Medical College, Mumbai.

“..... Examinees who wish to anticipate routine questions and to avoid long embarrassing silences would do well to read these pages..... important points have received appropriate emphasis..... The text is closely written and the amount of information provided is truly gross. Every line and word has to be remembered.....” —Book review (Part I) in Journal of the Indian Medical Association (JIMA).

“The monograph on Bedside Clinics in Medicine is very well written, studded with your long experience as clinical teacher. Such monograph was a long felt need. You have really done an excellent job. The monograph will be very well received not only by undergraduate and postgraduate students but by clinical teachers as well.” —Dr. A. P. Jain, Professor & Head, Department of Medicine, M. G. Institute of Medical Sciences, Sevagram, Maharashtra.

“I highly appreciate the efforts, hard work and sincerity in bringing up this publication in a beautifully designed manner with rich clinical material inside”. —Dr. S. B. Agarwal, Professor & Head, Department of Medicine, B. J. Medical College and Civil Hospital, Ahmedabad, Gujarat.

“..... It is a poetry on Clinical Medicine.” —Final year MBBS student, N.R.S. Medical College, Kolkata.

“..... I congratulate you for bringing out this book.....” —Dr. P. K. Rathor, P. G. trainee, M. K. C. G. Medical College, Berhampur, Orissa.

“..... The book is good and very useful to undergraduates & postgraduates in Medicine I shall continue to recommend your book..... congratulation to the author for his concentrated effort.” — Prof. (Capt.) G. Nagaiah, Professor & Head, Department of Medicine, Thanjavur Medical College & Hospitals, Tamilnadu.

“..... I have gone through this book and found it most suitable for the students. I will definitely recommend this book to the students.....” — Prof. (Dr.) D.K. Hazra, Director, Professor & Head, Department of Medicine, S.N. Medical College, Agra, Uttar Pradesh.

“..... Bedside Clinics in Medicine by Dr. Arup Kumar Kundu is a good book for undergraduate medical students. It is helpful for preparing for final MBBS Examination. Students can guess as what type of questions may be asked in the practicals.....” — Dr. B.T. Tukol, Professor of Medicine, K.M.C, Hubli, Karnataka.

“ is an excellent book for junior students in Medicine. This book, very simply written in a concised and rational manner will greatly help the undergraduate students to establish the foundation of Clinical Medicine with ease and confidence.” —**Dr. P. C. Bhattacharyya, Ex-Professor of Medicine, Gauhati Medical College, Assam.**

“ it is just excellent. It is an ideal companion for both undergraduates and post-graduates during exam time..... ” —**Dr. Neelakantan V. and Dr. N. Parvathi Sulochana, Sundarapuram, Coimbatore, Tamilnadu.**

“The second revised edition of Bedside Clinics in Medicine contains information at one place which postgraduates in Medicine aspire to assimilate in order to learn art and science of Medicine. Really an excellent job by Dr. Arup Kundu”. —**Dr. A. P. Jain, Professor & Head, Department of Medicine, M. G. Institute of Medical Sciences, Sevagram, Maharashtra.**

“..... It is so nice that all clinical problems are completed in so small a volume..... ”. —**Dr. K. Gandhi, Assistant Professor of Medicine, Thanjavur Medical College, Tamilnadu.**

“..... This is not a book but something more than that. This is self-explanative and could be one of the best books in the field of Clinical Medicine required for our students. The book is indispensable for not only undergraduates and postgraduate students but also for teachers and practitioners in Medicine as well. This book is a living Clinical Tutor”. —**Dr. Kiranmoy Mitra, Ex-Assoc. Professor & Head, Department of Medicine, Burdwan Medical College and Hospital, West Bengal.**

“Your fascinating presentation of long cases and short cases in Medicine has attended our presence towards your book..... ” —**R. Ravishankar and S. Gupta, Final year MBBS students, Thanjavur Medical College, Tamilnadu.**

“In keeping with the expanding horizon of medical sciences and the gallant gallops of a plethora of newly emerging methodologies, Dr. Arup. K. Kundu has very meticulously and ingenuously architected his master creation Bedside Clinics in Medicine, Parts I and II. A purist pedagogue, a massive mentor, and an exemplary exponent engrossed with an expansive professional expertise and competency, he has probed deeply into several cases along with history, diagnosis and management invariably in an easy-to-understand question-answer form. This lucidity, sometimes in a literary, and mostly in a highly scientific manners, has made this work an invaluable medical contribution-cum-anthology for undergraduate and postgraduate students. Moreover a beginner may find it a crutch to have a naive rendezvous into this area of medicine. The diagnostic procedures and methodologies are nicely delineated. This book will be a constant concise companion for all, students and teachers alike, in different Indian Universities and Medical Colleges as this treasure will elicit the clinical spirit of approach from a modest conventional way to highly sophisticated method.” —**Prof. (Dr.) C. R. Maity, Ex-Dean, Faculty of Medicine, Burdwan University; Principal, Burdwan Medical College, Burdwan, West Bengal (Ex-Director of Medical Education, WBMS, Government of West Bengal).**

“I happened to go through your book Bedside Clinics in Medicine It was nice and quite interesting..... I can recommend the book to my students as a ready reckoner..... ” —**Dr. V. Venugopal, Professor of Medicine, Perundurai Medical College, Tamilnadu.**

“The book Bedside Clinics in Medicine, Part I and II is comprehensive with wide coverage of all systems and attractively produced..... it is informative and beneficial not only to the UGs/PGs, but also to the physicians.” —**Col. A. S. Kasthuri, Professor and Head, Department of Medicine, AFMC, Pune, Maharashtra.**

“..... I found that there is a wealth of information in the book which is difficult to get from other books..... you must have gone through several journals and the whole of encyclopedia of medicine..... Congratulations.” —**Dr. K. Ramachandran, Visiting Professor of Radiology; MG University School of Medicine, Gandhi Nagar, Kottayam, Kerala.**

“..... I have been an ardent reader of both of your textbooks on clinical medicine, since the first day of my ward duty. To be very honest I have learnt more from your books than from.....” —Dr. Saif Omar, Internee, Katihar Medical College, Katihar, Bihar.

“..... many congratulations for writing an ultimate manuscript in medicine.....” —Dr. Kamaalchand M, P.G. trainee in Pediatrics, JNMC, Aligarh Muslim University, Aligarh, Uttar Pradesh.

“..... Bedside Clinics in Medicine, Part I & Part II is a Treasure Island” for students of Medicine both undergraduates & postgraduates..... The book has in fact satisfied the need of a long awaited reference book for examinee being complete by itself in all respects..... recommend this book strongly for students of medicine.....” —Prof. (Dr.) P. R. Nath Barbhuiya (Retd.), Professor & Head, Department of Medicine, Silchar Medical College & Hospital, Assam.

“..... Bedside Clinics in Medicine, Part I, which is highly informative, well-written with latest additions” —Dr. N. S. Neki, Associate Professor of Medicine, Government Medical College, Amritsar, Punjab.

“..... the joy of reading the book is so overwhelming that, by far, medicine has never seemed so pleasurable. It definitely provides so much of knowledge and information, that everytime I close the book after reading it, I do so with an extreme sense of happiness and confidence of knowing so much..... It is unequivocal opinion that the book is outstanding and entirely removes the need to study multiple books in clinical medicine..... For my final year exams..... the questions that were asked during discussion were entirely based upon the facts given in your book. I just had to quote them to be appreciated by the examiners. I wish to explain my gratitude as a student of medicine for your valuable contribution which is unfathomable” —Dr. Keerthana Karumbaiah K, Internee, Bangalore Medical College, Bangalore, Karnataka.

“..... this ‘made easy’, if assimilated properly by the students, will help to learn many aspects of medicine..... The chapters on radiological diagnosis and ECG interpretations will certainly help all concerned. Emergency tackling of different cases also, will help the young professionals..... The book is likely to be well accepted and the second edition of the book supports that expectation.” —Book review (Part II) in Journal of the Indian Medical Association (JIMA).

“..... its really the eighth wonder in the world. Looking back, recollecting my MD days, I repeat my words..... I owe you my MD.” —Dr. Pradeep Kumar Shenoy C, Ex-clinical fellow and registrar, Department of Rheumatology, Manipal Hospital, Bangalore.

“..... I am very happy to inform you that I have got selected for MD..... I owe my success to your book, and your in time valuable suggestion and advice. You have been my behind the scene teacher and educator and inspiration.....” —Dr. Bhushan Madke, student of Indira Gandhi Govt. Medical College, Nagpur, Maharashtra.

“..... You have donated breath to me.....” —Jagroop Singh, student of Guru Gobind Singh Medical College, Faridkot, Punjab.

“..... I feel it is a very good book and very informative.....” —Dr. George K. Chako (MD, PhD), PDH Hospital Group, Mumbai, Maharashtra.

“I am a final year medical student. I read your book on bedside clinical examination..... The book is really wonderful. Hats of sir.....” —J. Mahammad Sadiq, Government Medical College, Salem, Tamilnadu.

“..... I am highly impressed with clarity and concept of your book. The book is not only helpful for clinical exams but also for theory. The book is really a wonderful book.....” —Ramchandra Chaudhary, student of S.R.T.R Medical College, Ambajogai, Maharashtra.

"Sir, its really great whatever you write, I have read your book in final year to clear exam, that helped very much to pass final prof....." —Dr. Amit Sharma, (DM cardiology student), and passed MD (medicine) from KGMC, Lucknow, Uttar Pradesh.

"Bedside Clinics in Medicine by Dr. Arup Kundu is really nice to crash all the viva Q-A in your clinical viva.....also it helps to clear basics.....It teaches you how to think medicine"—in [http: / /medcosmosbaroda.blogspot.com/2008 09 01_archive.html](http://medcosmosbaroda.blogspot.com/2008_09_01_archive.html)

A website in Vietnam, recommended this book as only clinical medicine book from Asia—[www. ykhoavn. net/modules. php?name=Forums&file=viewtopic&p=7035-43k](http://www.ykhoavn.net/modules.php?name=Forums&file=viewtopic&p=7035-43k)

Website in China (Weifang Medical University, Shandong, China) recommends this book for their students— www.xmail.net/wfmustudents/medicalbooks.htm

www.rxpgonline.com

- "Medicine viva—by kundu; this comes in 2 volumes and is a must read stuff...."
- "Kundu is great for symptomatology and specific exam cases....."
- ".....go through a good clinical exam/cases book like Hutchinson and Kundu...."
- "Some important things you should know before entering clinics is given in Mcleods, Kundu clinical medicine. I think you can get these books and start off...."
- ".....It is wonderful for all category of students whether average or brilliant one. It teaches you the basic medicine....."

[mciforchina. blogspot.com / 2008/08/m-c-i-screening-t es t-books-syllabus. html-85k](http://mciforchina.blogspot.com/2008/08/m-c-i-screening-test-books-syllabus.html-85k)—recommended Bedside Clinics in Medicine, Part I & II for MCI screening test books for students passed MBBS from abroad.

www.medicalgeek.com (which book for clinical medicine?)

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- "For clinical exam I read everything from Kundu part 1 and 2.....never touched..... even Dr. Arup Kumar Kundu was our external examiner....."
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"..... also Arup Kundu if you want to dash answers in ward round cases. I would recommend Kundu as a must buy over and above one basic book like Hunter....." — www.aippg.net

"..... Favorite Medical Books: Guyton's Physiology & Kundu's Medicine_____ " in www.doctorshangout.ning.com/profile/DrMansi

"....Favorite medical books: Harrison's principle's of internal medicine, Kundu, de Gruchy, John Patten...." in www.doctorshangout.com/profile/PEUISHSUGATHAN

"I would like to meet: Harrison, Hutchison, Davidson, Kundu...." in [www.doctorshangout.com/profile / RAJESHARAHANT](http://www.doctorshangout.com/profile/RAJESHARAHANT).

"1 am a 4th year student at IMTU Dar Es Salaam Tanzania. I have seen one of book titled Bedside clinics in medicine. How can I get my copies..... "—hance Mdunye, IMTU, Dar Es Salaam, Tanzania.

".....It is a well known fact among medical professionals, that your book on bedside medicine has proved to be a holy gift to the generations of medical students in this part of the world—"—Apildev Neupane, final year medical student at Institute of Medicine, Kathmandu, Nepal.

..... and many others from every nook and corner of the country.

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(Perspectives

In 1816, Rene Theophile Hyacinthe Laennec invented the stethoscope — the single instrument that came to symbolize the bedside physician and the weapon used by the clinicians for almost last two centuries.

The term patient is derived from the Latin word 'patiens', or 'to suffer'. A 'patient' is one who feels sick physically or mentally and consults doctor for relief of his ailments. In clinical medicine, patients hold the key position. Naturally, it is the task of the physician that he should do the needful for the recovery of the patient.

To relieve the sufferings of the patients, a doctor should ascertain at first, the nature of illness from which the patient is suffering from. In other words, the doctor has to diagnose the existing disease. Physical diagnosis makes the bridge between the study of disease and the management of illness. Clinical diagnosis is an artistic science, based on three kinds of medical informations :

- (I) Communication with the patient (i.e., the history taking),
- (II) Physical examination, and
- (III) Investigations.

Clinical medicine deals with the first two parts of diagnosis i.e., 'clinical examination' of a patient consists of history taking as well as physical examination. These are the most important parts and it starts as soon as the patient comes to the doctor. At no point can **history taking** (medical events that have already taken place) be considered as 'wastage' of time; in most cases, the diagnosis can be made with history alone. History taking is an art, which a doctor should learn over years by repeated practice. Regarding history taking, it is my advice to the students of clinical medicine that they should try to disprove Ben Berston's comment, who told 'A funny thing happens to medical students on their way to becoming physicians; they forget how to hold a conversation'. While examining the patient, an experienced doctor not only 'sees' but also 'uses all of his senses' and gradually develops the keen 'power of observation' which is known as 'clinical eye' or 'clinical acumen'. These can not be acquired in a short-cut way and there is no rule of thumb in it. To learn this, one has to go to the bedside of the patients for years together, to make keen observations, to examine them thoroughly and sympathetically, and to try to 'associate' different observations and findings. Gradually the person develops the inherent 'knowledge of correlation' with the help of clinical eye. This is why, Hippocrates commented, 'A great part, I believe, of the art of medicine is the ability to observe. Leave nothing, combine contradictory observations and allow yourself enough time'. **An experienced clinician not only hears but listens, not only sees but observes, and not only touches but feels.** One, who has sharp clinical eye, becomes successful in life in the long run.

Traditionally, the process of **physical examination** (clinical state at that given time) is divided into four parts like looking (inspection), feeling (palpation), tapping (percussion), and listening (auscultation). From our student days we have learned to determine 'where is the lesion' as well as 'what is the lesion' derived by physical examination to arrive at a definitive diagnosis; this is also true to the students of this generation too. It should be realized that inspection / palpation / percussion / and specially auscultation are skills acquired over years of training. We know that our professional progenitors relied more on **power** of smelling (sweetish musty odour of hepatic coma) and tasting as well (the sweetness of urine in diabetes mellitus), but these facts have lost their importance in today's modern medicine. A sincere and meticulous physical examination combined with in-depth as well as sound knowledge of internal medicine would help the physician to determine the most probable cause / causes of the patient's present ailments. It is wise to consider the patients as living pages of books, and instead of examining from 'head to feet', I should rather advise students to examine patients from 'head to toes'.

Another important part of clinical medicine is doctor-patient relationship. It has got immense value in ultimate recovery of the patient. By the way of 'conversation' and physical examination, one sympathetic doctor gains confidence and wins trust of the patient. This should never be neglected. Whenever the

confidence of the patient is achieved, it becomes easier for the doctor to get his cooperation to examine the painful areas or even the diseased private parts of the body. In a non-cooperative patient, diagnosis may be much more difficult and delayed.

Investigations help the doctor to confirm the diagnosis made by clinical examination. Of course, clinical diagnosis by physical examination has limitations. No doubt, in many cases one has to take the help of newer and sophisticated investigations to arrive at a definitive diagnosis. Recently there is a boom in investigational facilities like polymerase chain reaction, ELISA test, ultrasound, CT scan, NMR scan, PET scan, SPECT scan where the patient is being fragmented into systems, organs, tissues, cells or even the smallest DNA, and very often these facilities erode the confidence of the clinicians concerned. Now-a-days, a large section of doctors feel that clinical examination is not at all essential and diagnosis can be made easily with the help of sophisticated investigations only. But this is not at all true. The laboratory should be used as an aid for confirmation of diagnosis and the final approach should be to 'treat the patient', not the laboratory reports. One confident doctor should never disregard his own clinical findings; moreover, newer investigations are often very costly and out of reach of most of the patients of a developing country like India. A long time back, Alvan R. Feinstein remarked, 'Clinical judgement depends not on knowledge of causes, mechanisms or names of disease but on a knowledge of patients. The background of clinical judgement is clinical experience; the things that clinicians have learned at the bedside is the care of sick people'.

In spite of these facts we can not ignore the importance of 'chemical' diagnosis and contribution of physics in medicine e.g., the discovery of X-ray by Wilhelm Roentgen or ECG by Willem Einthoven. So question arises, will this computer and sophisticated newer technology replace the physician's task of performing physical examination? The answer is simply 'no'. The computer necessarily deals in the cold literal aspects of words, whereas the doctors are trained to search for hidden meanings. To the question, 'is your pain continuing at the present moment?' The mechanical computer only records 'yes' even when the patient shows agony while responding. The doctor reads far more to the patient's response to 'yes'. Moreover, detailed questioning and physical examination frequently build the first emotional contact between the doctor and the patient. Can a computer be sympathetic to the patient regarding his illness? Or can a fascinating electronic gadget replace the excellence of Giant Clinicians like Batista Morgani, Leopold Auenbrugger, Rudolf Virchow, RTH Laennec, Sir William Osier, Adolph Kussmaul and Josef Francois Felix Babinski? A human brain created the computer, and thus a slave can neither demonstrate physical signs of the disease in a patient nor can interpret the complexities encountered to have a definitive diagnosis. So there is reason to believe that clinical medicine will ever persist as a basic diagnostic method and investigations will remain as 'complementary' to the clinician.

Unlike physics or chemistry, medicine is not a pure science. Half of what is true today will be proven to be incorrect in the next five years, and unfortunately we don't know which half that is going to be. The question arises whether medicine is a science, or an art, or an art based on science? The scientists say that medicine is sometimes considered a science, and sometimes an art, and the object of medical science is to study disease. I think medicine is an art based on science; medicine is supposed to be a scientific study whether its practice is an art, and does not consist of compounding pills and plasters only. Today, every senior doctor is shouting in every corner of this globe that the art of clinical medicine is dying in the present teaching set up with high-tech gadgets. I also think that they are mostly true; today, doctors are more powerful but more deaf. Medical students are advised to revive the dying art of medicine and to be good artists with sufficient scientific knowledge.

In the beginning of their clinical training, the students should also remember these golden words: **'there is no substitute for watchful eyes, alert ears, and tactful fingers in a logical mind'.**

CHAPTER I : LONG CASES

SCHEME OF CASE-TAKING

(A) Particulars (biodata) of the patient :

Bed No.-----

1. Name
2. Age
3. Sex
4. Religion
5. Occupation
6. Address
7. Date of admission
8. Date of examination

(B) The history or history proper :

1. Chief or presenting complaints
2. History of (H/O) present illness
3. Past history
4. Personal history
5. Family history
6. Treatment history
7. Psychological history
8. Menstrual and obstetric history in females

(C) Physical examination :

- I. GENERAL SURVEY —
 1. Level of consciousness; whether alert, oriented and co-operative
 2. Apparent age (corroborative or not)—Down's syndrome, thalassaemia and a pituitary dwarf look younger; and in progeria or precocious puberty, the patient looks older than his/her chronological age
 3. Decubitus (position of the patient in bed)
 4. Build or built (skeletal framework)—Average/dwarf/tall stature
 5. Nutrition (nourishment of the body)—Average/undemutrition/obese. Try to determine body mass index (BMI)
 6. Fades (facial appearance)
 7. Anaemia (mild, moderate, severe)—Probably 'pallor' is a better terminology
 8. Cyanosis (central or peripheral type)
 9. Jaundice (mild, moderate, severe)
 10. Neck vein (jugular venous engorgement, pressure and pulsation)
 11. Neck artery (carotid arteries)
 12. Lymph nodes (all over the body)
 13. Thyroid gland
 14. Clubbing
 15. Koilonychia
 16. Pulse (with all the points)
 17. Respiration (rate, rhythm, type, depth and breathing pattern)
 18. Temperature (record oral temperature)
 19. Blood pressure — Preferably both in supine (compulsory) and standing position. Measurements both in right and left arm are sometimes necessary
 20. Oedema
 21. Skin, hairs (including head to see alopecia) and nails
 22. Height and weight, arm span, upper segment-lower segment ratio (anthropometry)
 23. Any obvious deformity (of skull, spine, limbs, or swelling of abdomen)

24. General : any acute distress present or not
25. Handedness (right or left) with level of intelligence (average, low, high)

II. SYSTEMIC EXAMINATION —

1. Cardiovascular system (CVS)
2. Respiratory system
3. Gastrointestinal system (G.I. system) or alimentary system
4. Nervous system
5. Genitourinary system
6. Lymphoreticular system
7. Locomotor system (optional)

(D) Summary of the case (construct two paragraphs. First paragraph : short history in two or three sentences, and second paragraph : examination findings in brief with major abnormalities constructed in two or three sentences)

(E) Provisional diagnosis

(F) Differential diagnosis

(G) Relevant investigations (optional)

* In the history sheet, one may add 'points in favour of diagnosis' after the provisional diagnosis.

** In the systemic examination, one must write 'all' the systems, e.g., in a patient with mitral stenosis —first the CVS is dealt with in details and then the other systems are considered in brief.

*** Stand on the right side of the patient during examination.

THE HISTORY

1. CHIEF COMPLAINTS :

Major complaints with the duration should be written in the patient's own language in chronological order of their appearance.

2. H/O PRESENT ILLNESS :

- a) Expansion of the chief complaints in relation to their mode of onset (acute/subacute/insidious), progress (progressive/static/with exacerbations and remissions) and duration. Progress during stay in the hospital should be enquired into.
- b) Positive, plus **important negative points** (e.g., paralysis in neurological disorder or breathlessness in cardiovascular diseases).
- c) Use patient's own words and avoid scientific or medical terms as far as possible (joint pain is preferable term than arthritis).
- d) Do not put leading questions (i.e., questions suggesting their own answers) but many a time direct relevant question makes an essential component of sensitive history-taking.
- e) Record relevant associated symptoms.

D Generalities — Appetite, loss of weight, fatigue, sleep, bladder and bowel.

* Bladder and bowel habit may be included in personal history instead.

3. PAST HISTORY :

History of relevant past illness like,

- | | |
|--|---|
| a) Rheumatic fever. | j) 'H/O contact' with persons suffering from tuberculosis or any contagious disease. |
| b) Tuberculosis. | k) 'H/O exposure' (sexual) to STD. |
| c) Malaria. | l) Any illness which demanded 'blood transfusion' (e.g., accidents or any operation). |
| d) Kala-azar. | m) Childhood illness (e.g., eruptive fevers). |
| e) Jaundice. | n) Past hospital admissions. |
| f) STD (sexually transmitted diseases like gonorrhoea, syphilis, AIDS etc.). | o) Any other major medical or psychiatric illness in the past. |
| g) Systemic hypertension. | p) H/O travel (abroad or disease-prone areas). |
| h) Diabetes mellitus. | |
| i) Trauma or injury. | |

In pediatrics age group take the birth history (asphyxiated or not), H/O immunisation, past H/O any injury, umbilical sepsis or meningitis from the parents.

Few clinicians prefer to mention hypertension and diabetes mellitus within personal history.

*** Never comment, 'past history nothing significant', rather say 'the patient had no history suggestive of tuberculosis, jaundice..... and rheumatic fever'.

4. *PERSONAL HISTORY :*

, This includes H/O the patient, spouse and children only. Following points (e.g., personal details and lifestyle) should be noted carefully :

- a) Marital status with number of children.
- b) Occupation (nature and environment of job) and education; unemployment.
- c) Income (asked indirectly) and social (socio-economic) status.
- d) Addiction (tea, coffee, smoking, alcoholism, or substance abuse e.g., chewing tobacco, cannabis, heroin; try to estimate the amount of consumption of tobacco or alcohol).
- e) Dietary habit (for diagnosis of avitaminosis, malnutrition, obesity),
- f) History of contraception.
- g) Whether performing exercise regularly or not?
- h) High risk behaviour (e.g., IV drug abuse, multiple unprotected sexual exposure, homosexuality etc)—important in hepatitis B or C infection, AIDS and SBE.
- i) Domestic and marital relationship; hobbies and pets.

* Tobacco abuse : form (cigarette/biri), quantity and duration of exposure

5. *FAMILY HISTORY :*

This includes the parents, brothers, sisters, uncles (maternal or paternal), nephew etc. H/O tuberculosis (affected by contact), diabetes mellitus, systemic hypertension, ischaemic heart disease (IHD), bronchial asthma, eczema, haemophilia, thalassaemia, schizophrenia within the family should be enquired into. H/O similar type of illness (as the patient is suffering from) in the family should be asked for. Cause of death of the near relatives (parents) should be mentioned. H/O consanguineous marriage within the family should be recorded (autosomal recessive disorders e.g., homocystinuria may result from consanguineous matings). Family history is important in hereditary and communicable diseases (e.g., mumps, measles, chickenpox). Environmental factors should always be taken into account e.g., effects of passive smoking in a woman who is having a heavy-smoker husband. Lastly, the 'pedigree chart' (symbols used in construction of a family) may be drawn.

6. *TREATMENT HISTORY :*

- a) Treatment received so far, for the present illness.
- b) Any H/O drug allergy or reactions.
- c) Any surgical intervention or H/O accidents in significant past.
- d) Prolonged use of oral contraceptives (may precipitate CVA), penicillamine (used in Wilson's disease; may develop nephrotic syndrome) or vitamin C (may produce oxalate stone) etc.
- e) Blood transfusion—frequency, amount, type (e.g., whole blood, packed cell).
- f) Intake of NSAID (may produce acute gastric erosion, NSAID-induced asthma etc.).
- g) Regular user of oral contraceptives, vitamins, laxatives, sedatives or herbal remedies.
- h) Immunisation.
- i) Self-medication.

7. *PSYCHOLOGICAL HISTORY :*

Observe the 'mood' of the patient, i.e., whether anxiety, depression, irritability, euphoria, obsession, neurosis, depersonalisation are present or not. Peptic ulcer, bronchial asthma, irritable bowel syndrome are examples of psychosomatic disorder.

8. *MENSTRUAL AND OBSTETRIC HISTORY :*

■ These include the following :

- a) Menarche.
- b) Duration of the period.
- c) Quantity of blood loss (usually assessed by number of pads consumed or passage of clots).
- d) Dysmenorrhoea, amenorrhoea or other menstrual irregularities.
- e) Date of last menstrual period.
- f) Menopause, post-menopausal bleeding.
- g) Obstetric history—
 - (i) No of pregnancies
 - (ii) Outcome of pregnancies; H/O abortions, or carried to term; live birth (male/female)
 - (iii) Complications during pregnancy (e.g., hypertension, gestational diabetes mellitus)
 - (iv) Mode of delivery (vaginal, forceps. Caesarean)
 - (v) Last child birth

* Occupational history (exposure to dust/chemicals, working in a high altitude, coalmines and other business affairs) may be taken into account.

N.B.: While history-taking, full concentration is paid to the patient and the barriers of communications (foreign visitors, difficulty in hearing/vision, dysphasic/aphasic patient, emotionally disordered i.e., angry or abusive patient) may be overcome by information obtained from a relative. Catch the non-verbal clues (e.g., catch in breath, flushing, restlessness or changing eye contacts are signs of 'stress') in the patient. One must be very careful on highly sensitive topics in history-taking e.g., drug addiction, eating disorder, marital disharmony, sexual abuse or offending behaviour. Remember, in case of females, there might be some triggering influence of menstruation on migraine or pregnancy on heart failure.

During history-taking be sympathetic and show patience, and always try to be on the patient's level in all respect. The skills of history taking can be obtained and maintained satisfactorily only by practice.

ESSESI

CARDIOVASCULAR SYSTEM

The symptoms of CVS are mentioned below :

1. Breathlessness (dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea)
 2. Chest pain (angina pectoris, acute myocardial infarction or AMI, pericardial pain, aortic dissection)
 3. Palpitation
 4. Cough and expectoration
 5. Haemoptysis (expectoration of blood, or bloody sputum; seen in mitral stenosis)
 6. Dizziness and syncope; black-out
 7. Convulsions or past H/O neurodeficit
 8. Tiredness or fatigue (aortic stenosis, [5-blocker therapy)
 9. Swelling of feet (peripheral oedema) or facial puffiness
 10. Intermittent claudication or limb pain
 11. Squatting or cyanotic spells (cyanotic congenital heart diseases)
 12. Visual disturbance (retinal haemorrhage or embolism)
 13. G. I. symptoms like anorexia, nausea, vomiting (digitalis toxicity or protracted congestive cardiac failure [CCFD], pain abdomen (mesenteric embolism, tender hepatomegaly in CCF, inferior wall AMI)
 14. Renal symptoms like oliguria, nocturia (in CCF)
 15. Hoarseness of voice or dysphagia (left atrial enlargement due to mitral valve disease, or aortic aneurysm)
 16. Fever (SBE, tuberculous pericarditis, rheumatic activity, deep vein thrombosis, Dressler's syndrome)
 17. Fleeting joint pain, sore throat, subcutaneous nodules for rheumatic heart diseases
 18. General symptoms — Appetite, loss of weight (cardiac cachexia) or gain in weight (due to edema), fatigue, sleep, bladder and bowel
- * No. 1, 2, 3, 6 and 9 are cardinal symptoms in cardiology

Scheme of Examination

(A) Pulse :

1. Rate
2. Rhythm
3. Volume
4. Condition of the arterial wall
5. Comparison between two radial pulses
6. Radio-femoral delay
7. Any special character
8. Other peripheral pulses

* May add **blood pressure** here

(B) Examination of neck veins :

1. Engorged or not
2. If engorged,
 - (i) Pressure
 - (ii) Pulsation — Present or not
 - (iii) Hepato-jugular reflux — Present or not

(C) Examination of the heart :**I. INSPECTION :**

- a) Inside the precordium —
 - (i) Shape — Presence of any deformity or bulging.
 - (ii) Pulsation — Apical impulse; diffuse pulsation over precordium (see page 42).
 - (iii) Engorged superficial veins — Present or not (see page 436).
 - (iv) Polythelia (accessory nipple) — Often indicates underlying congenital heart disease, cardiomyopathy etc.
 - (v) Any scar mark (specially for previous cardiac operation), sinuses.
- b) Outside the precordium — Any pulsation present in.
 - (i) Aortic area.
 - (ii) Pulmonary area.
 - (iii) Parasternal areas (right and left).
 - (iv) Epigastrium.
 - (v) Suprasternal area.
 - (vi) Carotid pulsation.
 - (vii) Locomotor brachialis.
 - (viii) Back — Specially at the inferior angle of scapula in coarctation of aorta (Suzman's sign).
- c) Inspection of the BACK for scoliosis, kyphosis, gibbus, drooping of the shoulder, winging of the scapula; examine the skin for any sinus, ulcer, venous or arterial prominence, scar mark etc.

* Look for midline sternotomy scar for coronary artery by-pass surgery, left submammary scar for mitral valvotomy and infraclavicular scar for implantation of pacemaker.

II. PALPATION :

- a) Mitral area —
 - 1. Apex beat -
 - (i) Site, and
 - (ii) Character
 - 2. Palpable heart sound (M¹)
 - 3. Thrill
- b) Pulmonary area —
 - 1. Pulsation
 - 2. Palpable heart sound (P₂)
 - 3. Thrill
- c) Aortic area —
 - 1. Pulsation
 - 2. Palpable heart sound (A₂)
 - 3. Thrill
- d) Tricuspid area —
 - 1. Left parasternal heave
 - 2. Palpable heart sound (T₁)
 - 3. Thrill
- e) Direction of venous blood flow (in engorged superficial veins).
- f) Thrill in carotid arteries (carotid shudder).
- g) Epigastric or suprasternal pulsation, or any pulsation over the back.
- h) Palpable pericardial rub.
- i) Tracheal tug.

* See page 418 for different areas of heart, pages 30 and 42 for thrill, page 416 for palpation of heart sounds and parasternal heave.

III. PERCUSSION : Not done**IV. AUSCULTATION :**

- a) Cardiac rate
- b) Rhythm
- c) Mitral area —
 - 1. Heart sounds
 - 2. Murmur (in detail)
 - 3. Adventitious sounds — Opening snap, ejection click, splitting of heart sound, S₃ or S₄
- d) Pulmonary area — As in mitral area
- e) Aortic area — As in mitral area

- f) Tricuspid area — As in mitral area
- g) Auscultation over neo-aortic area
- h) Auscultation over carotid arteries for bruit (after holding the breath)
- i) Venous hum at root of neck (e.g., severe anaemia)
- j) Pericardial rub

* It is better to write 'audible' in case of heart sounds which are not produced in that area. Thus, in pulmonary hypertension S_2 (P_2) is loud but S_j is audible in pulmonary area.

** Always examine female patients in the presence of a female attendant.

*** Before doing any clinical procedure or manoeuvre (e.g., palpation of apex beat) always take permission of the patient and explain briefly what you are going to do.

**** Always think of some common disease by the 'law of probability'; if you give the provisional diagnosis of a rare disease, you are rarely correct. Remember the dictum : Uncommon presentations of common diseases are more common than common presentations of uncommon diseases.

Case 1

MITRAL STENOSIS

What is your diagnosis ?

Give the provisional diagnosis (P/D).

It is a case of mitral stenosis (MS) of rheumatic origin without any evidence of congestive cardiac failure (CCF) and the patient is in sinus rhythm at present.

What is your case ?

One should describe the summary of the case without mentioning the P/D directly, e.g.,

Sunita Devi, 26 years female patient presented with gradually increasing breathlessness for last 10 months, and cough with haemoptysis for last 1 month. The breathlessness was gradually progressive, exertional, non-seasonal and NYHA (New York Heart Association) grade II in severity. Cough was not associated with rise of temperature. She had no H/O palpitation, chest pain, swelling of legs, syncope, squatting (to exclude cyanotic congenital heart diseases), pain in legs during walking or any neurodeficit. Two weeks back, she experienced acute respiratory distress at middle of night due to paroxysmal nocturnal dyspnoea (PND). She was neither hypertensive nor diabetic; there was no H/O contact with tuberculosis and no H/O exposure. She had an uneventful H/O child birth, 4 years back. She had definite H/O rheumatic fever in her childhood which affected mainly the big joints like knee, ankle and elbow following an attack of sore throat. The arthritis was fleeting in nature and not associated with any skin lesion or chorea.

On examination, there is no anaemia; pulse is 78/minute, low volume, regular with all the peripheral pulses palpable equally; BP is 100/70 mm of Hg. in supine position and the neck veins are neither engorged nor pulsatile. There is no deformity of precordium (precordial bulging indicates early onset and longer duration cardiac diseases). On palpation, the apex beat is present in left 5th intercostal space (ICS) 1/2" inside the left midclavicular line (MCL) and is tapping in character (i.e., S_j is palpable). A diastolic thrill is palpable in the mitral area which is best felt in left lateral position and in full expiration, and there is absence of left parasternal heave. On auscultation of the mitral area the 1st heart sound (S_1) is short, sharp, accentuated, and the 2nd heart sound (S_2) is audible. Opening snap (OS) is heard just after S_2 . There is a low-pitched mid-diastolic rumbling murmur with presystolic accentuation of grade IV intensity in the mitral area without any radiation (use the term 'radiation' or direction of selective propagation' but it is better to avoid the term conduction of murmur). The murmur is best audible at cardiac apex with the bell of stethoscope (placed lightly), in left lateral position, at the height of expiration and after doing mild exercise. There is absence of split, click, rub or murmur over the other areas. Examination of the respiratory system revealed no abnormality (no adventitious sound, no hydrothorax etc.) and there is absence of hepatosplenomegaly or ascites.

* Features of right ventricular hypertrophy (RVH) and pulmonary hypertension are absent in this patient.

** Past H/O rheumatic fever, hypertension, diabetes mellitus, tuberculosis (and syphilis) are important in all CVS cases.

*** A big summary is elaborated here for the sake of better understanding of the case history of MS.

Give the summary of the case :

Give the short case history from the question discussed above.

Provisional diagnosis in valvular heart diseases :

1. Anatomical — Mitral, tricuspid, aortic, pulmonary.

2. Structural abnormality—Stenosis, regurgitation.
3. Aetiological — Rheumatic, congenital.
4. Complications — Pulmonary hypertension, CCF, subacute bacterial endocarditis (SBE) etc.
5. Rhythm — Sinus rhythm or dysrhythmia (e.g., irregularly irregular rhythm due to atrial fibrillation).

Some clinicians prefer to add NYHA classification in the provisional diagnosis (see page 24).

Why it is a case of mitral stenosis ?

It is a case of mitral stenosis because of the following :

(A) Symptoms (from the history) :

- a) Breathlessness or effort intolerance for last 10 months.
- b) Cough with haemoptysis from time to time for 1 month.
- c) Attacks of paroxysmal nocturnal dyspnoea since last 2 weeks.
- d) Past H/O rheumatic fever (tell about the symptoms).

(B) Signs :

- a) Pulse - 78/minute, LOW VOLUME, REGULAR, no radio-radial or radio-femoral delay, all the peripheral pulses are palpable, no abnormality in the arterial wall or no special character.
- b) BP - Low (or normal).
- c) Apex beat - Normal in position and tapping in character. There is presence of diastolic thrill, best palpable in left lateral position and at the height of expiration. S₁ is palpable.
- d) Auscultation (of the mitral area) :
 S₁ - Short, sharp and accentuated (loud and snapping S₁ — a very important clinical clue).
 S₂ - Normal.
 Opening snap — Audible, just after S₂.
 Murmur - Describe the classical murmur (as described in summary).
 Other areas - No murmur.

* ADD FEATURES OF PULMONARY HYPERTENSION. IF PRESENT.

** Auscultatory cadence of murmur and heart sounds of MS are as follows: **ffout** (presystolic murmur ending in loud S₁) — **ta** (S₂) — **ta** (opening snap) — **rrrou** (mid-diastolic murmur).

*** 'Usually' the apical impulse in an established MS is diffuse and formed by right ventricle. There is a diastolic thrill at apex, best palpable at left lateral position. There is sustained left parasternal heave as a result of RVH from pulmonary hypertension.

If no **past H/O rheumatic fever present** :

Rheumatic fever is the commonest aetiology of MS and it is true in other valvular heart diseases. 60% of the MS patients do not give definite H/O rheumatic fever, still one should consider rheumatic fever as the probable aetiology. In that case it is better to say in P/D as 'a case of MS probably of rheumatic origin'.

Importance of past and family history in CVS :

Past history

1. Rheumatic fever
2. Cyanotic spells with H/O squatting
3. Recurrent respiratory tract infection
4. Any murmur or cardiac lesion detected at school
5. Hypertension, diabetes mellitus, IHD
6. Thyrotoxicosis

Family history

1. Hypertension
2. Congenital heart disease
3. Rheumatic heart disease
4. Ischaemic heart disease (IHD)
5. Obesity, diabetes, dyslipidaemia
6. Sudden (cardiac) death

How to diagnose congenital / early onset cardiac disease ?

1. History with ailments since birth/childhood with special reference to squatting.
2. Bulged precordium.
3. Clubbing with cyanosis plus polycythemia.
4. May be associated with dwarfism.

What is 'precordium' ?

It is the anterior chest wall which overlies the heart. In health, it is slightly convex and associated with a smooth contour. The different deformities are :

- a) Bulging - Early onset and longer duration cardiac diseases (e.g., VSD, rheumatic heart diseases), pericardial effusion, scoliosis, mediastinal tumours, left-sided pleural effusion.

- b) Flattening - Fibrosis of the lung, congenital deformity of chest, adherent pericarditis.

What is mitral fades?

It is the pinkish purple patches on cheeks. As low cardiac output in MS produces vasoconstriction, peripheral cyanosis is often seen in lips, tip of the nose and cheeks. Occasionally along with these, malar flush is seen due to vasodilatation (vascular stasis) in malar area. All these features constitute 'mitral facies' and is rarely seen in India.

Causes of malar flush :

1. High altitude.
2. Myxoedema.
3. Mitral stenosis.
4. Cushing's syndrome.
5. Thyrotoxicosis.
6. Chronic alcoholism.
7. Polycythemia.
8. Carcinoid syndrome.
9. Menopausal syndrome
10. Pheochromocytoma.

How loud S_j in MS can be explained?

Due to persistent diastolic gradient across the mitral valve, the valve cusps remain wide open throughout the diastole and as soon as the ventricular systole starts, the widely opened mitral valve cusps close rapidly, giving rise to loud S_j (think of the sound produced when a door is closed from a short distance or from widely opened position, and it is obvious that closing the door from a long distance will produce more sound).

Factors influencing the intensity of S_j :

1. Position of valve cusps (mitral or tricuspid) at the onset of ventricular systole :
 - a) Wide open valve cusps — MS or TS, hyperdynamic circulatory states (produce loud S_j).
 - b) Valve cusps remaining close to each other or can not close completely (e.g., MI or TI) —The S_j becomes muffled e.g., in myocarditis, myocardial infarction, heart failure, MI or TI.
2. PR interval (in ECG) — S_j is loud if PR interval is short, and S_j becomes muffled if PR interval is prolonged.
3. Pliability of the valve cusps e.g., calcification results in loss of pliability, and the heart sound becomes muffled or faint.
4. Ventricular muscle mass (booming S_j is heard in patients with systemic hypertension leading to LVH).
5. Heart rate — In tachycardia, S_j tends to be loud as a result of short PR interval.
6. Intervening media between heart (valve cusps) and the stethoscope e.g., emphysema, pericardial effusion or obesity diminishes the intensity of S_j.

* Normally the S₁ is dull and prolonged (lubb in 'lubb-dup'), and best audible at the apex. S₁ indicates the beginning of ventricular systole and is produced due to closure of atrioventricular valves. It has two components : mitral (Mj) and tricuspid (Tj).

* S_j coincides with carotid pulsation while S₂ comes after the pulsation. During auscultation of heart sounds in different areas, always palpate right carotid artery with the left thumb.

Table 1 : Variations in intensity of S_j

Loud S_j	Soft or muffled S_j
<ol style="list-style-type: none"> 1. Mitral stenosis, tricuspid stenosis 2. Sinus tachycardia due to any cause 3. Hyperkinetic circulation e.g., severe anaemia, pyrexia, exercise, pregnancy, thyrotoxicosis, Paget's disease, beriberi 4. Short PR interval, e.g., tachycardia, nodal rhythm 5. Normally in children 	<ol style="list-style-type: none"> 1. Mitral incompetence, tricuspid incompetence 2. Prolonged PR interval (e.g., 1° heart block), bradycardias 3. Right or left ventricular dysfunction 4. Calcified mitral/tricuspid valve 5. Obesity, emphysema, thick chest wall, pericardial effusion
Varying intensity of S_j	Splitting of S_j
<ol style="list-style-type: none"> 1. Atrial fibrillation 2. Ventricular tachycardia 3. Complete heart block (cannon sound) 4. Multiple extrasystoles 	<ol style="list-style-type: none"> 1. Wide splitting : complete RBBB, left ventricular pacing, Ebstein's anomaly 2. Reversed splitting : complete LBBB, right ventricular pacing, severe MS, left atrial myxoma

Muffled S₂ in MS :

When MS is associated with :

1. Mitral incompetence (MI) or severe aortic incompetence (AI).
2. Mitral valve calcification.
3. Active rheumatic carditis (due to prolonged PR interval).
4. Digitalis overdose (due to prolonged PR interval).
5. Acute myocardial infarction.
6. Left atrial failure (LAF).
7. Atrial fibrillation (produces varying intensity of S₂).
8. Rotation of heart due to gross right ventricular hypertrophy (RVH) and consequently right ventricle is forming the apex.
9. Emphysema, obesity, thick chest wall (transmission of S₂ is hampered).

Comment on apex beat in MS :

Usually the apex is in normal position. If the apex goes outward, think of RVH (from pulmonary hypertension) and if it is downward plus outward, think of LVH (i.e., search for associated MI, AI, AS, systemic hypertension). Normally, the apex is tapping (palpable SJ in character and may be diffuse (due to RVH). LVH is not a feature of isolated MS.

How do you clinically assess the 'severity' of MS ?

Features of severe MS are :

1. Narrow S₂-OS gap (the smaller the gap, the more severe is the MS), and
2. Longer duration of mid-diastolic murmur.

N.B. : Remember, it is the duration and not the intensity, which assesses the severity of MS. Other signs of severity are : symptomatic patient, low volume pulse, RVH and pulmonary hypertension in ECG, and cardiomegaly in chest X-ray.

Recognition of pliable mitral valve cusps :

1. Short, sharp, accentuated S₂, and
2. Presence of opening snap.

* If the valve cusps are non-pliable (i.e., in mitral valve calcification), S₂ will be muffled and the opening snap will not be heard.

Absent presystolic accentuation in MS :

1. Atrial fibrillation,
2. Left atrial failure,
3. Big left atrial thrombus.

Why the murmur is mid-diastolic in MS ?

The interval between closing of the aortic valve (S₂) and opening of the mitral valve is known as 'isovolumetric relaxation' period. Diastolic murmur of MS is heard as soon as the blood flows from the left atrium to left ventricle after opening of the mitral valve (i.e., in the mid-diastole). There is no blood flow in the isovolumetric relaxation period and this is why the murmur of MS is mid-diastolic, not early diastolic.

The last part of ventricular diastole (last rapid filling phase) actually coincides with atrial systole. Atrial systole increases the blood flow across the stenotic valve and thus, there is accentuation of the diastolic murmur (presystolic accentuation).

What is an opening snap (mitral) ?

(A) Opening snap is produced due to bellowing down of the closed mitral valve cusps at the onset of ventricular diastole (i.e., mitral valve will open just now but has not opened yet). It is :

- a) Sharp and high-pitched.
- b) Best heard with the diaphragm of stethoscope.
- c) Best heard after expiration with the patient in standing position.
- d) Loudest in between the apex beat and the left sternal border.
- e) Present in early diastole (at the end of isovolumetric relaxation period).
- f) It may be the loudest sound in the cardiac cycle.

(B) It indicates :

- a) MS is organic.

- b) Valve cusps are pliable.
- c) Significant MS.
- d) High left atrioventricular pressure gradient.
- e) Severe AI, gross MI, gross pulmonary hypertension, atrial fibrillation, left atrial failure or SBE (subacute bacterial endocarditis) is absent.
- f) Diminishing S₂-OS gap indicates increasing tightening of stenosis (severe MS).
- g) MS is readily amenable to surgery.

* Opening snap is also audible in tricuspid stenosis, ASD, VSD, PDA, posterior cuspal type of MI.

** To differentiate OS from splitting of S₂, ask the patient to stand. As standing reduces the venous return, the S₂-OS gap increases and A₂-P₂ gap of S₂ decreases on assuming standing position (dynamic auscultation).

Aetiology of MS :

MS is 'almost always' or invariably of **rheumatic** in origin. Rarely the congenital form ('parachute' mitral valve—all chordae inserted into a single left ventricular papillary muscle) may produce MS, or very rarely it may arise from severe mitral annular calcification, carcinoid syndrome, collagen vascular disease (SLE), endomyocardial fibrosis or mucopolysaccharidosis. MS results from shortening of chordae tendineae and fusion of commissures.

* IN CLINICAL PRACTICE IT IS SAID THAT THE COMMONEST AETIOLOGY OF MS IS RHEUMATIC, THE SECOND IS RHEUMATIC AND THE THIRD IS ALSO RHEUMATIC.

** Rheumatic MS is much more common in females.

Pathological types of MS :

1. Button hole, 2. Fish mouth, and 3. Funnel type.

* Mitral valve apparatus is composed of 1. valve leaflets, 2. valve annulus, 3. chordae tendineae 4. papillary muscles, and 5. myocardium (the papillary muscles are attached here).

Common symptoms of MS :

Dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea (PND), palpitation, fatigability, haemoptysis, recurrent bronchitis, cough, chest pain, right hypochondrial pain (tender hepatomegaly), and symptoms related to complications.

- * 1. Symptoms due to low cardiac output—fatigability,
- 2. Symptoms due to left atrial failure (i.e., pulmonary congestion)—dyspnoea, orthopnoea, PND, cough, haemoptysis, recurrent bronchitis.
- 3. Symptoms due to RVF—swelling of legs (oedema), pain in right hypochondrium.
- 4. Symptoms due to arrhythmia—palpitation.

** Chest pain in MS is due to 1) RVH, 2) coincidental coronary atherosclerosis, or 3) coronary embolism.

Lutembacher's syndrome :

Atrial septal defect (ASD) plus MS (of rheumatic origin).

Changes noticeable when atrial fibrillation develops in MS :

1. Ventricular rate (heart rate) is about 100-150/minute.
2. **Irregularly irregular pulse with varying volume, and a pulse deficit > 10.**
3. Varying intensity of S, (classical loudness of S_j is lost); opening snap is not heard.
4. Absent a-wave in the neck veins or JVP.
5. **Presystolic accentuation of the diastolic murmur disappears;** only mid diastolic rumble persists.
6. Congestive cardiac failure may precipitate and embolic manifestations may appear (CVA, intermittent claudication, pain abdomen etc).

* *Aetiology of atrial fibrillation* are rheumatic (commonly MS), ischaemic, hypertensive, thyrotoxicosis, cardiomyopathy, myocarditis, pericarditis, idiopathic or 'lone' atrial fibrillation (in elderly persons without any demonstrable heart disease), congenital (especially ASD) and sinoatrial disease.

** In atrial fibrillation, the atria usually fire impulses at a rate of 350-600/minute.

Changes occurring after left atrial failure in MS producing pulmonary oedema :

1. S, becomes muffled,
2. Opening snap is not heard.

3. Presystolic component of the murmur disappears, and
4. Presence of bilateral basal crepitations (fine).

Which chamber of heart fails first in MS ?

The left atrium. Chamber involvement in MS : LA → RV → RA.

Which ventricle fails first in MS ?

The right ventricle (**left ventricle does not fail in MS as mitral valve lies proximal to it**)

* In the presence of left ventricular hypertrophy or failure in MS, think of associated valvular diseases like MI, AI, AS, or systemic hypertension, ischaemic heart disease, cardiomyopathy.

What is tight or severe MS ?

Normal cross-sectional area of mitral valve orifice is 4-6 cm² (average 5 cm²). Classification in relation to dimension of mitral valve orifice is : Minimal — >2.5 cm², mild—1.6 to 2.5 cm², moderate — 1 to 1.5 cm², and severe (tight/critical) — <1 cm².

What is juvenile mitral stenosis :

This type of MS is usually found in India. In India, MS develops early in contrast to the west where the patient experience disability in the 4th decade. The criteria for diagnosis are :

- | | |
|----------------------------------|-------------------------------------|
| 1. Occurs below 18 years of age. | 4. Valve calcification is uncommon. |
| 2. Pin-point mitral valve. | 5. Needs immediate operation. |
| 3. Atrial fibrillation is rare. | 6. Common in South-East Asia. |

Haemoptysis in MS : explanation :

The possible mechanisms are —

1. Pulmonary apoplexy — Due to rupture of thin-walled, dilated bronchial veins or pulmonary veins (frank haemoptysis) resulting from sudden rise in LA pressure.
2. Winter bronchitis (chronic bronchitis) — Blood-streaked mucoid sputum.
3. Acute pulmonary oedema — Profuse, pinkish, frothy sputum is produced due to rupture of capillaries into the alveoli.
4. Pulmonary infarction — Frank haemoptysis.
5. Overdose of anticoagulant therapy (rare) — Often required for embolic manifestation.
6. Pulmonary haemosiderosis — Rare.

Most significant finding in your case :

The mid-diastolic rumbling murmur.

What is damped MS ?

Development of pulmonary hypertension diminishes the cardiac output (throttle effect) and results in temporary symptom-free period (period of illusion). Often the mid-diastolic murmur is not audible and thus known as silent MS.

What is intermittent claudication ?

This is a cramp-like pain, tightness and numbness (due to ischaemia) felt commonly in calves, thighs or buttocks on walking a certain distance, and characteristically relieved by taking rest. The 'claudication' (Latin meaning 'to limp') suggests peripheral vascular disease and the most important risk factor is smoking. The common causes are,

1. Buerger's disease (thromboangiitis obliterans; young heavy-smoker males).
2. Atheroma or severe atherosclerosis of lower limb arteries; Leriche's syndrome (embolism at branching of common iliac artery i.e., claudication of thigh + impotence).
3. Coarctation of aorta.
4. Arteritis.
5. Lumbar canal stenosis (patient often stoops forward during walking to reduce symptoms. If he continues to walk, paraesthesia in feet and even foot drop may develop).
6. Venous claudication (bursting pain on walking, previous H/O DVT), and
7. Over-exertion.

Palpation of peripheral pulses in legs, i.e., arteria dorsalis pedis shows diminution in the volume of pulse in 1, 2, 3 and 4. In 5, 6 and 7, peripheral pulses remain normal but in 5, ankle jerk may be diminished or absent.

- Vascular claudication (No. 1, 2, 3, 4) : Cold legs, pallor, feeble pulse are common. Common risk factors are diabetes, smoking, hypertension and hypercholesterolaemia.

- Neurogenic claudication (No. 5 or cauda equina compression) : Paraesthesia, limb weakness, diminished ankle jerk with normal pulse.

Differential diagnosis (D/D) of mitral diastolic murmur :

1. Carey-Coombs murmur — Soft mid-diastolic murmur of active rheumatic valvulitis, localised to the apex. The murmur varies in intensity from day to day and usually disappears after the acute attack. There is absence of loud S₁, opening snap and diastolic thrill. Oedema of the mitral valve cusps produce obstruction and gives rise to diastolic murmur.

2. Austin Flint murmur — It is a functional low-pitched, mid-diastolic rumbling murmur produced in a patient with severe AI. When the aortic regurgitant jet of blood impinges on the anterior mitral leaflet (thereby stenosing the mitral orifice relatively), a murmur (actually the murmur is due to both antegrade flow of blood from left atrium and regurgitant jet from AI) is audible with the following features :

- Mid-diastolic murmur with absence of presystolic component; heard in the mitral area.
- No thrill.
- S₁ is not loud.
- Absence of opening snap.
- Commonly found in syphilitic AI.
- Isometric hand-grip increases the murmur.
- Presence of AI murmur in the aortic area with features of LVH, plus
- Peripheral signs of AI (read the section on 'Aortic incompetence').

3. Functional mid-diastolic murmur — Found in increased left-to-right shunt in ventricular septal defect (VSD), atrial septal defect (ASD), or patent ductus arteriosus (PDA). Increased flow through the normal mitral valve also occurs in severe MI. There is absence of diastolic thrill, opening snap and presystolic accentuation but S₃ may be present.

4. Left atrial myxoma—

- Alteration of physical signs (murmur) with change of posture; H/O postural syncope.
- Tumour 'plop'—a sound heard as the pedunculated tumour comes to a halt.
- Constitutional symptoms like fever, weight loss, anaemia, arthralgia, rash; high ESR.
- Embolitic episodes.
- 2-D echocardiography is diagnostic.

5. Tricuspid stenosis — Mid-diastolic murmur with presystolic accentuation which is loudest at lower left sternal edge. Murmur increases at the height of inspiration. Features of RVH are never present.

6. Ball valve thrombus — The thrombus usually floats in the left atrium and obstructs the mitral orifice in diastole. Echocardiography is essential for final diagnosis.

7. Conducted murmur of AI — Early diastolic, soft blowing; no thrill, no opening snap. Peripheral signs of AI are present.

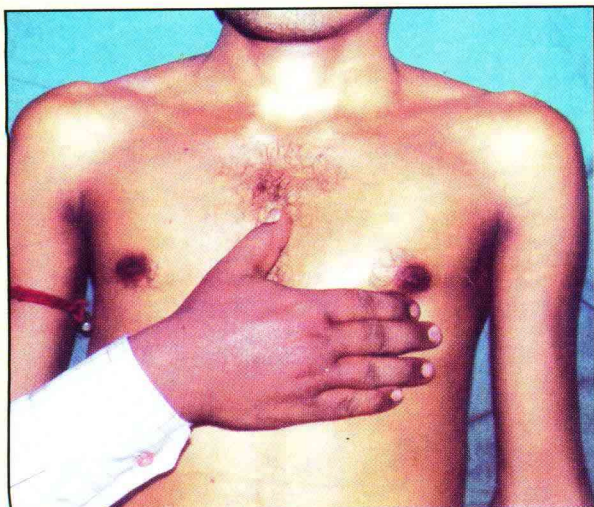
8. Cor triatriatum — A correctable malformation of heart where an abnormal fibromuscular diaphragm separates the left atrium into two chambers. Echocardiography and cardiac catheterisation are diagnostic.

* Closest D/D of MS are left atrial myxoma and ball valve thrombus in the left atrium.

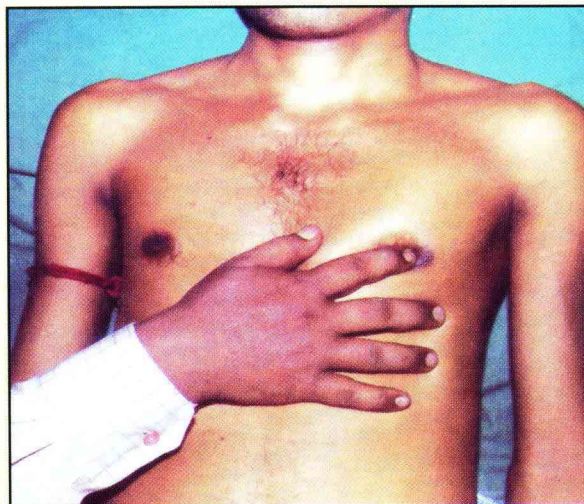
Describe the classical attack of paroxysmal nocturnal dyspnoea (PND) :

It is the sudden and dramatic development of acute dyspnoea occurring in the early hours of night, i.e., nearly 30 minutes to 2 hours after retiring to bed. The patient is awakened from sleep with a feeling of apprehension, intense suffocation and choking sensation. He sits upright gasping in the bed with the legs hanging by the side of the bed (to reduce left atrial pressure by gravitational pooling) or rushes to an open window in the hope that cool fresh air will relieve him. Dyspnoea progresses with profuse sweating. These are accompanied by a dry, repetitive cough which is due to '**acute interstitial oedema**' resulting from pulmonary venous hypertension, when there is no collection of fluid within the alveoli. The attack may subside spontaneously within 30 minutes but often progresses to '**acute pulmonary oedema**' which may end fatally. The cough becomes productive with profuse, watery, pinkish and frothy sputum due to acute pulmonary oedema (collection of fluid within the alveoli).

PND signifies the earliest symptom of acute left-sided heart failure (due to LAF or LVF). Acute dyspnoea with wheeze and repetitive productive cough are features of '**cardiac asthma**'. Actually 'cardiac asthma' is closely related to PND and is characterised by wheezing due to bronchospasm which is most prominent at night. Acute pulmonary oedema is the severe form of cardiac asthma due to marked increase in pulmonary capillary pressure leading to alveolar oedema.



Palpation of cardiac impulse (supine position)



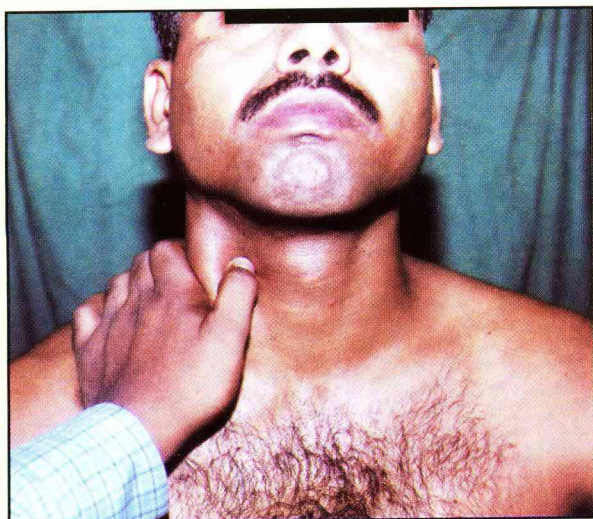
Localizing apex beat with the fingers (supine position)



Palpating apex beat for character (left lateral position)



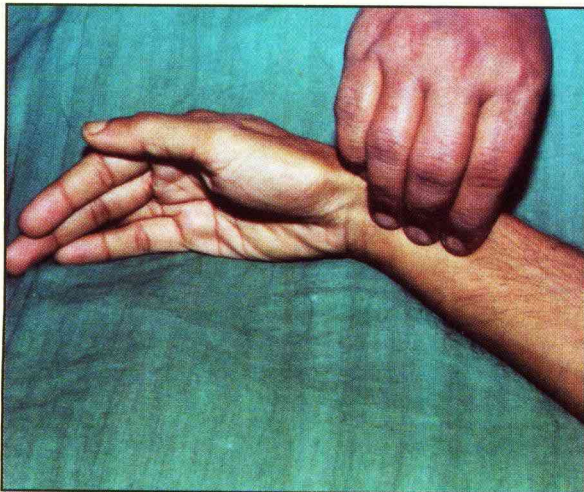
Auscultation of mitral area (especially for MS) : use bell while patient in left lateral position



Palpation of carotid artery (carotid pulse)



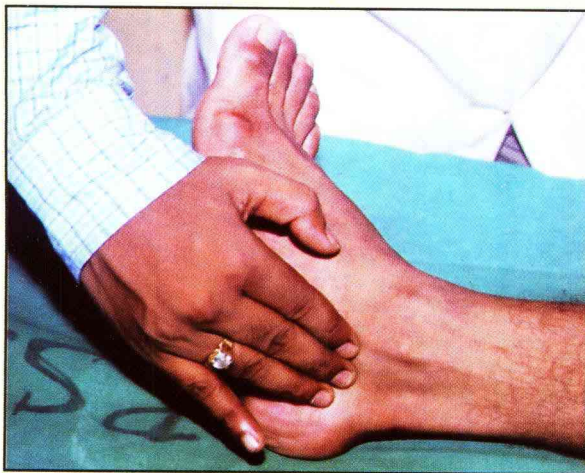
Auscultation of aortic area : use diaphragm while patient is sitting and leaning forward



Palpation of **radial artery** ('the pulse')



Palpation of **arteria dorsalis pedis** (pedal pulse)



Palpation of **posterior tibial artery**



Examination for **radio-femoral delay**



Examination of **popliteal artery**



Elicitation of **water-hammer pulse** (collapsing radial pulse)

Clinical examination at the time of an attack of PND reveals :

- a) Severe dyspnoea and orthopnoea; tachypnoea associated with air hunger and recruitment of accessory muscles.
- b) Anxious and pale (ashen-grey pallor) patient with profuse sweating; cold extremities.
- c) Central cyanosis.
- d) Pulse — Tachycardia; feeble pulse.
- e) BP — May be high (as hypertension is the commonest cause of PND).
- f) JVP — Raised.
- g) Heart — S₃ gallop rhythm; signs of underlying heart disease.
- h) Lungs — Vesicular breath sound with prolonged expiration; bubbling crepitations +++ (predominantly basal), and scattered rhonchi + present.

* Acute pulmonary oedema is the phenomenon of "cough, cough, cough — spit, spit, spit".

**** So, causes of LAF and LVF are the basic causes for paroxysmal nocturnal dyspnoea or acute pulmonary oedema.** PND is a symptom of decompensation of the left heart.

*** 'Left side' of the heart encompasses the functional unit of left atrium, left ventricle, mitral valve and aortic valve; and the 'right side' of the heart is used for the term encompassing right atrium, right ventricle, tricuspid valve and pulmonary valve.

****In recumbency, the rapid increase in venous return leads to pulmonary congestion and the left-sided chambers can not cope up with this increased inflow in MS. Breathlessness in PND is due to inappropriate awareness of respiratory tract and firing of 'J' receptors in lung.

Why dyspnoea occurs in MS ?

It is known that abnormally elevated LA-LV pressure gradient is the haemodynamic hallmark of MS. The events in MS go like this : TLA pressure → Tpulmonary venous pressure → Tpulmonary capillary wedge pressure → icompliance of lung → dyspnoea (exertional).

Protective mechanisms to prevent pulmonary oedema in MS :

As the disease progresses to chronic mitral stenosis, there is development of :

1. High pulmonary vascular resistance (pulmonary hypertension),
2. Capillary-alveolar-interstitial barrier, and
3. Broncho-pulmonary venous shunts.

Thus, it is clear that a recent onset MS patient may die of LAF (acute pulmonary oedema) and an old or chronic patient dies of RVF (from pulmonary hypertension). An old case of MS usually does not suffer from PND because of the development of protective mechanisms mentioned above.

Causes of 'non-cardiogenic' pulmonary oedema (loosely termed as 'ARDS') :

Pulmonary oedema is of two types ; cardiogenic and non-cardiogenic. The causes of non-cardiogenic type are—

1. Diffuse pulmonary infections (viral, bacterial, fungal, pneumocystis jiroveci).
2. Inhalation of toxins and irritants (phosgene, chlorine, high concentration of O₂).
3. Aspiration of vomitus, water (near drowning), acid/alkali poisoning, kerosine poisoning.
4. Narcotic (heroin, morphine, methadone, dextropropoxyphene) and non-narcotic (nitrofurantoin) overdose/effects.
5. Severe sepsis, gram negative septicaemia, shock, major trauma, uraemia.
6. Acute haemorrhagic pancreatitis, amniotic fluid/fat/air embolism, fulminant hepatic failure, insecticide poisoning, snake bite, high altitude, eclampsia.
7. Miscellaneous — Cardio-pulmonary bypass, major blood transfusion reaction, anaphylaxis (bee, wasp, snake venom, crab), major burns, head injury, SLE, DIC, lymphangitis carcinomatosa.

* In severe cases, pink and frothy sputum comes out through the nose. **Remember, pneumonia (exudate) is not pulmonary oedema (transudate).**

** 'Cardiogenic' pulmonary oedema — results from left-sided heart failure (LAF/LVF), acute myocardial infarction, cardiac dysrhythmias, acute pulmonary thromboembolism.

*** Right heart catheterisation by Swan-Ganz catheter allows differentiation of two types. **Cardiogenic**—engorged neck veins, TCVP (central venous pressure) and tPCWP (pulmonary capillary wedge pressure); **non-cardiogenic**—normal left-sided atrial or ventricular pressure, neck veins are not engorged, and with normal CVP and PCWP.

Why the dyspnoea is usually nocturnal in MS ?

Probable theories are :

1. Venous return increases in recumbency.
2. Mobilisation of oedema fluid from extravascular to intravascular compartment on lying.
3. Adrenergic drive is reduced during sleep.
4. Heart rate increases during rapid eye movement (REM) sleep.
5. Reduction of vital capacity in supine position.
6. Elevation of left atrial pressure and fall in PaO_2 during sleep.

Complications of MS :

1. Acute left atrial failure and acute pulmonary oedema.
2. Pulmonary hypertension.
3. Right ventricular failure (RVF/CCF).
4. Atrial fibrillation (AF), atrial flutter, ventricular or atrial premature beats.
5. Embolic manifestations (20%) due to AF (from dislodged clots formed in the LA),
 - a) Cerebral - 60% (cerebral infarction leading to hemiplegia)
 - b) Peripheral arteries - 30% (aorta—Leriche's syndrome)
 - c) Visceral - 10%
6. Haemoptysis.
7. Infective endocarditis — Very rare (more common in milder form of MS than in severe form); endocarditis is common in 'regurgitation' than in 'stenosis'.
8. Recurrent broncho-pulmonary infections.
9. Complications due to giant left atrium (LA)—
 - a) Hoarseness of voice — Due to compression of left recurrent laryngeal nerve by enlarged LA (Ortner's syndrome); very rare.
 - b) Dysphagia — Due to oesophageal compression.
 - c) Clot present in a giant LA may give rise to embolic manifestations even in sinus rhythm.
10. Jaundice, cardiac cirrhosis.
11. Sudden death may be caused by ball valve thrombus where the LA outlet is blocked by large pedunculated thrombus.

* Complications not related to severity of MS are atrial fibrillation, embolic manifestations and SBE.

How do you like to investigate a case of MS ?

- (A) Chest X-ray (PA view) —
- a) Slight increase in transverse diameter of heart (due to RVH).
 - b) 'MITRALISATION' of heart means straightening of the left border of heart and is due to (from above downwards) :
 - 1) Aortic knuckle — Small (due to low cardiac output).
 - 2) Convexity due to dilated pulmonary artery (due to pulmonary hypertension).
 - 3) Left atrial appendage — becomes prominent and produces a convexity.
 - 4) Left border of left ventricle — NO CHANGE.
 - c) Double contour of the right border of heart (shadow within shadow) — The outer and upper border is due to LA, and the inner and lower border is due to RA enlargement.
 - d) Evidence of 'pulmonary hypertension' — Dilated pulmonary arteries at hilum with peripheral pruning (i.e., peripherally pulmonary arteries taper sharply).
 - e) Dilatation of upper lobe pulmonary veins (earliest X-ray feature of pulmonary venous hypertension).
 - f) Fan-shaped opacity ("bats'-wing" appearance from parahilar region to periphery indicates pulmonary oedema).
 - g) Kerley's B lines — Fine, dense horizontal lines at the base of the lung due to distension of interlobular septa and lymphatics, with oedema.
 - h) Mitral valve calcification — Best seen in fluoroscopy.
 - i) Elevation of left upper lobe bronchus which becomes horizontal (due to LA enlargement).
 - j) Multiple small opacities from pulmonary haemosiderosis (subjects who had multiple haemoptysis may show haemosiderin deposits in the lungs) and parenchymal ossification—rare.

- (B) Chest X-ray (RAO view) — Right anterior oblique (RAO) view with barium-filled oesophagus is done. Oesophagus is pushed or curved backward (sickling of oesophagus) by the enlarged LA.
- (C) Chest X-ray (lateral view)—Obliteration of retrosternal space in left lateral view with slight increase in transverse diameter of heart (PA view) indicates RVH. -Uprturned' apex (in PA view) may result in boot-shaped configuration due to RVH.
- (D) Electrocardiography (ECG) — In early stages, the ECG may be normal.
 - a) LA enlargement — Wide and notched P-wave (P-mitrale)
 - b) RVH
 - c) 'f-waves replacing P-waves, if atrial fibrillation develops
- (E) Blood examination (for active rheumatic carditis) —
 - a) Total and differential count (TC and DC) b) ESR c) ASO titre
- * All are raised in the presence of active carditis.
- (F) Echocardiography (M-Mode and 2-D) to see chamber enlargement, valve pathology, valve movement, mitral orifice etc (in MS there are thickened immobile cusps, reduced valve area, LA enlargement and reduced rate of diastolic filling of LV).
- (G) Cardiac catheterisation — To determine the haemodynamic status of the patient (pressure measurement, oxygen saturation in blood samples)—pressure gradient between LA and LV is determined.
- (H) Doppler study — To know the functional status, speed and direction of blood flow in the heart chambers; to record pressure gradient across mitral valve.

**** Echocardiography confirms the diagnosis of valvular heart disease.**

Rationality of nervous system examination in MS i.e., in CVS cases :

1. Hemiplegia or monoplegia may occur in a patient with MS (commonly with atrial fibrillation).
2. Sydenham's chorea.
3. Fundoscopy — Roth spot, hypertensive retinopathy.

Rationality of respiratory system examination in MS i.e., in CVS cases :

1. Respiration (tachypnoea, orthopnoea).
2. Hydrothorax (from CCF).
3. Crepitations at lung bases (due to left-sided heart failure).

Rationality of G. I. system examination in MS i.e., in CVS cases :

1. Liver — Soft and tender liver with mild enlargement due to CCF.
2. Spleen — In the presence of SBE, spleen may be palpable.
3. Ascites may develop from CCF.
4. Mesenteric embolism.

Rationality of skeletal system examination in cardiac disorders :

1. Build—
 - a) Tall — Marfan's syndrome (MI or AI).
 - b) Stunted growth — Turner's syndrome (coarctation of aorta).
2. Kyphoscoliosis (shifting of apex beat, deformity of precordium).
3. Polydactyly or syndactyly (congenital heart disease).
4. Cubitus valgus (Turner's syndrome may be associated with coarctation of aorta).
5. Fingerisation of thumb with upper limb dysplasia (Holt-Oram syndrome with ASD).
6. Rheumatoid arthritis or ankylosing spondylitis (MI or AI); rhumatic arthritis (carditis).

* Heart diseases may be congenital or acquired. The external features for *congenital heart diseases* are : cyanosis, clubbing, polycythemia, webbing of neck, hypertelorism, syndactyly, low set ears, cryptorchidism etc. In *acquired heart diseases*, search for stigmata of rheumatic fever (arthritis, subcutaneous nodules etc.), infective endocarditis (anaemia, clubbing, splenomegaly, Osier's node etc.) and coronary heart diseases (xanthelasma, arcus senilis, obesity, nicotine staining, ear lobe creases etc.).

Important points in general survey in CVS :

1. **Decubitus** - May be propped-up; orthopnoea.
2. Build - Dwarfism in cyanotic congenital heart diseases; tall in Marfan's syndrome.
3. Nutrition - Cardiac cachexia in severe chronic heart failure.

4. **Anaemia** - Present in SBE (remember, anaemia can aggravate all heart diseases).
5. **Cyanosis** - Cyanotic congenital heart diseases, acute pulmonary oedema.
6. **Clubbing** - Cyanotic congenital heart disease, SBE, Eisenmenger's syndrome.
7. **Pulse** - Very important clue to diagnosis in different cardiovascular disorders.
8. **BP** - Must be seen in all patients: of immense value in diagnosis.
9. Respiration - Tachypnoea, hyperventilation.
10. Temperature - Elevated in SBE, tuberculous pericarditis.
11. **Oedema** - CCF, constrictive pericarditis, pericardial effusion, tricuspid valve disease.
12. **Neck vein** - Reflects central venous pressure; found in RVF.
13. Thyroid - One should search for thyrotoxic features in all CVS disorders.
14. Jaundice - Rare; due to congestive hepatomegaly from CCF, or cardiac cirrhosis.
15. Skin - Osier's node, ptechial haemorrhages, rheumatic subcutaneous nodules.
16. Skeletal - Kyphoscoliosis, cubitus valgus, polydactyly.

Treatment of MS :

Mild MS may need no treatment. The different modalities of treatment are :

- a) Drugs (medical treatment).
- b) Percutaneous mitral balloon valvuloplasty (treatment of choice).
- c) Valvotomy or commissurotomy — Closed and open (closed type is preferred). Open valvotomy needs open heart surgery with cardio-pulmonary by-pass.
- d) Mitral valve reconstruction including annuloplasty.
- e) Valve replacement or prosthesis (Starr-Edwards ball valve or Bjork-Shiley disc valve).

Indications of valvotomy :

1. Progressive symptomatic deterioration inspite of medical treatment.
2. MS with complications (as mentioned earlier) e.g., haemoptysis.
3. Asymptomatic patients with a single attack of thromboembolic manifestation.
4. MS with pregnancy where previous pregnancy was symptomatic.
5. Mitral valve orifice $<1 \text{ cm}^2$.

Contraindications of closed valvotomy :

- | | |
|---|---|
| 1. MS with significant MI. | 5. Extremely tight stenosis. |
| 2. MS with left atrial thrombus. | 6. Mitral valve distorted by previous operation |
| 3. Valvular calcification. | (i.e., restenosis cases). |
| 4. Presence of active rheumatic carditis. | |

Actually, these are indications for open valvotomy.

N.B. : Approximately 85% patients develop restenosis at 10 years after mitral valvotomy. Loud S, and OS persist after mitral valve prosthesis.

Criteria for mitral valvuloplasty :

1. Significant symptomatic MS.
2. Pure MS (may have trivial MI).
3. Left atrium is free of clots.
4. Valve and sub-valvular apparatus are free of calcification.

Treatment done in MS with MI:

Mitral valve replacement (prosthesis). Mechanical prosthesis like Starr-Edwards valve or Bjork-Shiley valve, or porcine bioprosthesis may be used. Prosthesis is done in grossly damaged valve and sub-valvular structures, and in presence of MI. Artificial valves may work for more than 20 years.

Medical treatment in MS :

1. Treatment of CCF by restriction of physical activity, salt-restricted diet, diuretics and digoxin (available as 0.25 mg tablet).
2. Antibiotic prophylaxis against infective endocarditis is advised though it is uncommon in pure MS (see the section on 'Aortic stenosis'). Secondary prevention of acute rheumatic fever is done.
3. Anticoagulation in the presence of atrial fibrillation or big left atrial clot to reduce the risk of systemic embolism.

4. Treatment of atrial fibrillation with digoxin, (3-blockers or rate-limiting calcium channel blockers (singly or in combination).
5. Management of complications like haemoptysis, acute pulmonary oedema are done accordingly.

MS with pregnancy : outline of management :

Rheumatic heart disease accounts for 90% of all cardiac diseases in pregnancy. Of these, MS is the commonest lesion. The principles of management are,

1. Schedule treatment of MS. Note : functional degradation (e.g., patient deteriorates from grade I to grade II NYHA classification) occurs in pregnancy due to increase in cardiac output.
2. Increased bed rest.
3. Adequate nutrition and correction of anaemia.
4. Therapeutic abortion—in selected cases e.g., with H/O repeated heart failure before conception, controlled heart failure in 1st trimester.
5. Mitral valvotomy—in symptomatic tight MS, in midtrimester.
6. Ideally all other cases should go into labour with scrupulous monitoring and not allowing the patient to strain in 2nd stage of labour.
7. Caesarean section may be performed where indicated.
8. Patients are advised to restrict the number of children to one or two.

Metallic mitral valve prosthesis in MS—auscultatory findings :

1. A metallic sound is audible in mitral area which coincides with S₁.
2. Normal S₂.
3. A metallic opening snap.

Prosthetic valves are of two types :

- Metallic valve—Starr-Edwards, Bjork-Shiley, St. Judes valve
- Tissue valve (human—homograft; porcine or bovine—xenograft)—Carpentier-Edwards, Ionescu-Shiley. Hancock porcine valve.

Complications of prosthetic valve :

Thromboembolism (needs anticoagulation), infective endocarditis, valve failure/leaking/obstruction by thrombosis or calcification/perforation/rupture, microangiopathic haemolytic anaemia etc.

N.B. : One should not use abbreviations like MS, MI, PH, BP etc; use the full term e.g., mitral stenosis, mitral incompetence, pulmonary hypertension, blood pressure respectively. In this book, abbreviations are used just to avoid repetition of long words. One should search for signs of active rheumatic carditis in all valvular heart diseases. Before stating the P/D as MS, one must be sure that there are no other valvular lesion like AI, MI or AS. If MS is associated with pulmonary hypertension, some changes develop in the palpatory and auscultatory findings — carefully search for them: often they are missed (read the section on 'Mitral incompetence' for the features of pulmonary hypertension).

Case 2

MITRAL INCOMPETENCE

What is your diagnosis ?

This is a case of organic mitral incompetence (regurgitation) of rheumatic origin with features of congestive cardiac failure and pulmonary hypertension, and the patient is in sinus rhythm at present.

What is your case ?

Build up the summary. The symptoms are almost similar to that of MS. Symptomatology consists of exertional dyspnoea, orthopnoea, PND, fatigue, recurrent respiratory tract infections, palpitation, oedema feet due to CCF or features of SBE (fever, embolic episodes etc.).

Palpitation is due to.

1. LVH, and / or
2. Atrial fibrillation (palpitation in MS is due to RVH or atrial fibrillation).

Why it is a case of mitral incompetence (MI) ?

It is a case of MI because of the presence of :

(A) Symptoms (from the history) :

- a) Exertional dyspnoea, and
 - b) Palpitation for last 6 months.
- (plus past H/O rheumatic fever).

(B) Signs :

- a) Decubitus - Propped-up.
- b) Pulse - Rate is 110/minute; otherwise normal arterial pulse (rarely the pulse is jerky or of good volume); describe all the points.
- c) JVP - Engorged and pulsatile (CCF).
- d) Oedema - Bipedal and pitting in nature (CCF).
- e) Precordium - Bulged.
- f) Epigastric pulsation - Present (RVH).
- g) Apex - Present in left 7th ICS, 1/2" outside the left MCL (LVH); hyperdynamic in character; systolic thrill is present in apical area which is best palpable in left lateral position and at the height of expiration.
- h) Left parasternal heave - Present (RVH).
- i) Palpation of pulmonary area - Diastolic shock (palpable P₂) present,
- j) Auscultation—
 - (i) Mitral area -
 - S₁ — Soft
 - S₂ — Audible
 - S₃ or S₄—Not audible at present

There is a high-pitched, soft blowing pansystolic murmur of Grade IV intensity which is best heard with the diaphragm of stethoscope, in left lateral position of the patient and at the height of expiration. The murmur is radiated towards the left axilla and inferior angle of left scapula (**hallmark of diagnosis**)

- (ii) Pulmonary area - Ejection systolic murmur (different from pansystolic murmur at apex) with loud P₂.
- (iii) Other areas - Within normal limit, i.e., S₁ and S₂ are audible, and there is no murmur, split, click or rub.
- [k] G. I. tract —
 - (i) Liver—Mild (enlargement), soft and tender hepatomegaly (CCF).
 - (ii) Spleen—Not palpable.
 - (iii) Ascites—Absent.
- l) Respiratory system —
 - (i) Breath sound - Vesicular, rhonchi—occasionally present, crepitations - nil.
 - (ii) Hydrothorax—Not present.
- m) Nervous system—Within normal limits.]

* Actually, these are features of MI with pulmonary hypertension and CCF.

** 'Rocking motion over the precordium' in each cardiac cycle in MI may be due to retraction of the left ventricle as well as expansion of the left atrium during systole.

Features of LVH :

Apex goes down and outwards.

Features of RVH :

1. Apex goes outwards.
2. Left parasternal heave.
3. Epigastric pulsation.

* Cardiac apex remains down and outwards in combined LVH + RVH.

Cardinal signs of LVF (left heart failure / left-sided failure) :

Three cardinal signs are—

1. Gallop rhythm (addition of S₃ or S₄, along with S₁ and S₂ produces '**triple rhythm**'. Triple rhythm with sinus tachycardia is known as **gallop rhythm** — so called because of its resemblance with the cadence produced by galloping of horses). So, *presence of gallop rhythm indicates heart failure*.
2. Fine crepitations at lung bases (i.e., basal crepitations).
3. Pulsus alternans.

* Sometimes, there are associated Cheyne-Stokes breathing, tachypnoea, central cyanosis, LVH and signs of underlying disease. The patient usually c/o breathlessness (including orthopnoea and PND), fatigue and mental confusion. Remember, pulmonary oedema is a feature of left-sided heart failure.

**** Gallop** is of two types :

1. Quadruple gallop — Four distinct heart sounds due to audibility of S₃ and S₄ separately, and
2. Summation gallop — The extra sound is produced as a result of superimposition of S₃ on S₄ (i.e., three audible sounds in one cardiac cycle).

*** Ventricular gallop S₁, S₂, S₃; atrial gallop S₄, S₁, S₂

Cardinal signs of RVF (right heart failure / right-sided failure / CCF) :

Three cardinal signs are —

1. Engorged and pulsatile neck veins.
2. Mildly enlarged, soft, tender liver.
3. Dependent bipedal oedema (pitting).

* Sometimes there are associated cardiomegaly, peripheral cyanosis, pulsatile liver (rare), pansystolic murmur of TI and right ventricular gallop rhythm. The patient c/o breathlessness, cough, oedema, oliguria, G. I. tract disturbances (e.g., anorexia, nausea, vomiting, right hypochondriac pain due to stretching of hepatic capsule) and swelling of abdomen (ascites). Cardiac cachexia (due to T tumour necrosis factor, T basal metabolic rate, anorexia, nausea, vomiting) is not uncommon.

Precipitating causes of cardiac failure in a patient of valvular heart disease :

- Severe anaemia, pregnancy, respiratory tract infections.
- Tachyarrhythmias, infective endocarditis, pulmonary thromboembolism, thyrotoxicosis.
- Ongoing rheumatic activity.

What is apex beat ?

It is the lowermost and outermost part of the precordium where a DEFINITE (most important, and not necessarily the maximum) thrust can be felt.

When the same cardiac pulsation is seen, it is called **apical impulse**. 'Cardiac impulse' is a better terminology than apical impulse because any pulsation 'seen' over the precordium may not correspond with the apex beat detected by palpation. If the apical impulse is not visible in supine position of the patient (best visible from the right side of the patient by a tangential view), the patient is turned to left lateral position when the apical impulse may be visible somewhere near the left anterior axillary line. Causes of non-visibility of apical impulse are same as causes of non-palpable apex (see below).

What is the significance of apex beat ?

It is an important parameter by which one can assess the enlargement of heart (in the absence of push-pull effect of lung) and the activity of myocardium.

How to localise the apex ?

1. The patient should lie flat or supine. Place your whole flat of right palm over the precordium (always stand on the right side of the patient). Try to localise the definite thrust (not necessarily the maximum) with outstretched fingers. If a definite thrust is palpable, confirm it by pulp of index or middle finger, and locate it by counting the ribs; look, how far it is from the left midclavicular line (inside or outside).
2. If the apex beat is not palpable in dorsal decubitus position, ask the patient to sit and lean forward. Again try to locate it.
3. If the apex is not palpable even in sitting position, place your right palm over the right side of the chest (to detect the presence of dextrocardia or dextroversion).
4. If still not palpable — Say that 'the apex could not be localised properly'.

* Remember, the heart has some mobility within the chest and thus never turn the patient to left lateral position for localisation of the apex beat (in left lateral position, apex shifts about 1-2 cm towards the left). **Normal position of the apex beat is in the left 5th ICS, 1/2" inside the left MCL (adult).**

In palpating apex, why the finger is lifted in systole when the ventricle is contracting ?

This is due to the complex rotatory movement of the heart in systole (vortex like movement), one manifestation of which is the forward movement of apex (the rotatory movement is due to the complex spiral arrangement of ventricular muscles).

ises of non-palpable apex ?

1. Apex lying behind a rib.
2. Obesity or thick chest wall.
3. Pendular breast in elderly female.
4. Emphysema (COPD).
5. Pleural effusion (left).
6. Pneumothorax (left)
7. Constrictive pericarditis.
8. Pericardial effusion.
9. Acute myocardial infarction.
10. Deformity of the chest (gross kyphoscoliosis)
11. Thickened pleura (left).
12. Heart failure.
13. Heart may be somewhere else (dextrocardia).

Describe different characters of the apex beat :

The normal apex beat is early systolic outward thrust located at a point not more than 2-3 cm in diameter. The different types are :

1. **HYPERDYNAMIC** (volume overload apex) —It means 'forceful and ill sustained' impulse found in conditions with diastolic overload of the left ventricle like MI, AI, VSD, PDA etc. As there is no obstruction to the blood pumped from the left ventricle, the apex remains ill sustained.
2. **HEAVING** (pressure overload apex) — It is a 'forceful and well sustained' impulse found in conditions with systolic overload of the left ventricle like AS, systemic hypertension, coarctation of aorta, obstructive cardiomyopathy etc. As there is some obstruction in left ventricular out-flow tract, the apex becomes well sustained.
3. **TAPPING** (small area) or **SLAPPING** (diffuse)— When the accentuated first heart sound is palpable in MS or tachycardia due to any cause, it is known as tapping apex beat (remember, heart sounds are not palpable in health). The palpating finger is not lifted and the S₁ is felt as distinct palpable shock.
4. **HYPOKINETIC** — The thrust felt by the hand is minimal. It is found in acute myocardial infarction, pericardial effusion, constrictive pericarditis, myxoedema, peripheral circulatory failure.
5. **NORMAL** — Situated in the left 5th ICS, 1 /2" inside the left MCL which is a brief gentle tap not exceeding a 25-paisa coin size, not much forceful but usually palpable with certainty. In children, the apex beat is located in the left 4th ICS and in lean-tall persons, it is usually present in the left 6th ICS.

* There may be double kicking apex beat in IHSS (idiopathic hypertrophic subaortic stenosis or hypertrophic cardiomyopathy) and ventricular aneurysm.

** Regarding the **character of the apex beat**, some clinicians prefer the terms 'right ventricular (RV)' or 'left ventricular (LV)' type. In **LV type** the apex goes downwards and outwards as well as there is a larger impulse (usually more than a 25-paisa coin), and the impulse is maximal at the apex; if there is a left parasternal impulse, it is asynchronous with the apex beat. Whereas in **RV type**, the apex goes outwards as well as there is production of left parasternal heave, and both are synchronous; in RV type, the cardiac impulse is maximal over the lower left sternal border.

*** While palpating for pulsations, e.g., apex beat — use the pulp of the fingers; for thrills—use the base of the fingers; and for parasternal heaves—use the base of the hand (i.e., thenar and hypothenar eminences).

Different positions of heart :

- Levocardia — Cardiac apex on left side of chest (situs solitus).
- Dextrocardia — Cardiac apex on right side of chest.
- Mesocardia — Cardiac apex over the centre of chest.
- Dextrocardia with situs inversus (heart per se does not show any abnormality).
- Isolated dextrocardia — Dextrocardia without situs inversus; almost always have cardiac malformations.
- Isolated levocardia — Cardiac apex on left side with viscera rotated (partial or totally) to opposite side.
- Dextroversion — Always acquired; cardiac apex is on right side due to extracardiac causes like left-sided pleural effusion/pneumothorax, or right-sided fibrosis/collapse of lung (i.e., push-pull effects). Remember, trachea is moved to the right side too.

* 'Situs inversus' is the rotation of abdominal /thoracic organs to opposite site, either totally (totalis) or partially (partialis).

Clinical features of pulmonary hypertension :

Normal pulmonary artery pressure is 25 / 10 mm of Hg. Pulmonary hypertension is said to be present when mean pulmonary artery pressure is > 25 mm of Hg at rest or > 30 mm of Hg during exercise.

(A) Symptoms—

1. Breathlessness (often severe; exertional).
2. Easy fatigability.
3. Dizziness.
4. Syncope (due to low fixed stroke volume).
5. Chest pain (angina pectoris).
6. Cough; haemoptysis.

(B) Signs—

1. Pulse — Low volume.
2. Neck veins — a-wave is prominent.
3. Central cyanosis—in right-to-left shunt, lung diseases (COPD, interstitial lung disease).
4. a) **Inspection—**
 - (i) Visible pulmonary artery pulsation in left 2nd ICS.
 - (ii) Epigastric pulsation.
- b) **Palpation—**
 - (i) Apex goes outwards.
 - (ii) P_2 — Diastolic shock, i.e., P_2 is palpable.
 - (iii) Left parasternal heave.
 - (iv) Pulsation of pulmonary artery may be felt in upper left parasternal area.
- c) **Percussion—**Left 2nd ICS may be dull on percussion.
- d) **Auscultation (pulmonary area) —** The **classical sequence of events** are,
 - (i) S₁ — Audible.
 - (ii) Pulmonary ejection click.
 - (iii) Ejection systolic murmur (due to relative obstruction at pulmonary orifice).
 - (iv) Close splitting of S₂ with loud P₂.
 - (v) Graham Steell murmur — A high-pitched early diastolic decrescendo murmur due to functional pulmonary incompetence (pulmonary valve ring dilatation).
 - (vi) Right-sided S₃ (right ventricular gallop) — Heard at the lower left sternal border.

Among these features, the ejection systolic murmur with palpable as well as audible loud P_2 are commonly found. Remember, (i) Epigastric pulsation, (ii) Left parasternal heave, and (iii) Outward apex are features of RVH. RVH is usually present in a case of pulmonary hypertension of some duration.

- Chest X-ray- Bulging right and left pulmonary arteries at hilum, peripheral pruning of large pulmonary arteries, enlargement of RV and RA of heart.
- ECG - RV strain or hypertrophy, and RA enlargement.
- Echocardiography - RV enlargement with paradoxical motion of interventricular septum.
- Pulmonary function test, lung scan and pulmonary angiography in appropriate cases].

* For demonstration of 'Signs' anywhere in clinical medicine, always start from general survey.

** Unless otherwise mentioned, pulmonary hypertension means pulmonary arterial hypertension. As soon as pulmonary hypertension develops from valvular heart disease, it protects the patient from pulmonary oedema.

Causes of pulmonary hypertension :

Pulmonary hypertension is basically of two types : Primary pulmonary hypertension (No. 6 below) and secondary pulmonary hypertension (No. 1 to 7, except No. 6)

1. Passive — From left-sided heart diseases like MS, MI, AI, AS.
2. Hyperkinetic (left-to-right shunt) — ASD, VSD, PDA.
3. Vasoconstrictive (hypoxic) — Chronic cor pulmonale.
4. Obstructive — Pulmonary thromboembolism (acute cor pulmonale).
5. Obliterative — SLE, systemic sclerosis, polyarteritis nodosa.
6. Neurohumoral or idiopathic — Primary pulmonary hypertension (common in females in 3rd and 4th decades).
7. Miscellaneous — Sleep apnoea, Pickwickian syndrome, drugs like aminorex fumarate.

Gradation of dyspnoea :

Functional gradation of heart disease by New York Heart Association— '**NYHA classification**':

Grade I : No limitation of activities with ordinary physical work (activity), i.e., asymptomatic though the patient is suffering from organic heart disease (there is no grade 0).

Grade II : No limitation under resting condition but symptoms appear on ordinary activity.

Grade III : Limitation of activities on mild exertion or less than ordinary physical activity.

Grade IV : Limitation of activities at rest, restricting the person to bed or chair.

* Remember, this functional classification refers not only to dyspnoea but to symptoms like fatigue, palpitation, angina pectoris etc. This is actually a *classification of cardiac disability*. Dyspnoea due to cardiovascular diseases should always be quantified by this classification.

Aetiology of MI:

1. **Rheumatic** (commonest).
2. Functional Associated with LVH (due to dilatation of mitral ring) as found in systemic hypertension, aortic valvular diseases (AS, AI). IHD, myocarditis, cardiomyopathy or acute rheumatic carditis.
3. Mitral valve prolapse syndrome (MVPS).
4. Dysfunction of papillary muscle or chordae tendineae (due to IHD i.e., ischaemic heart disease).
5. Acute myocardial infarction (due to rupture of papillary muscle).
6. Traumatic — During mitral valvotomy.
7. Infective endocarditis.
8. Collagen vascular diseases Marfan's syndrome, SLE (Libman-Sacks endocarditis), rheumatoid arthritis, ankylosing spondylitis etc.
9. Dilated cardiomyopathy.
10. Congenital.

* Three most common causes of MI according to order of frequency are rheumatic. MVPS and IHD.

Causes of acute MI:

1. Acute myocardial infarction (papillary muscle dysfunction/or rupture of chordae tendineae).
2. Infective endocarditis (rupture of valve cusps or chordae tendineae).
3. Trauma in the chest.
4. Acute rheumatic fever.
5. Cardiac surgery.

What is mitral valve prolapse syndrome (MVPS) ?

The other names of this disease are systolic click-murmur syndrome, Barlow's syndrome, floppy-valve syndrome. There is excessive mitral leaflet tissue which is commonly involved with myxomatous degeneration. MVPS is frequently associated with Marfan's syndrome, Ehlers-Danlos syndrome and osteogenesis imperfecta. It is common in young females. The patient is usually asymptomatic or may present with palpitation or chest pain. A mid- or late systolic click with high-pitched late systolic murmur best heard at the apex is the classical auscultatory finding. Treatment is done by propranolol for chest pain and arrhythmia, antibiotic prophylaxis for SBE, antiplatelets (e.g., aspirin) for prevention of transient ischaemic attacks (TLA), and valve replacement by prosthesis for permanent remedy.

What is seagull murmur ?

If a patient develops SBE over MI, the character of MI murmur changes with a musical quality called seagull murmur. Here the ruptured chordae tendineae acts as the string of the musical instrument. Seagull murmur is also known as 'cooing dove murmur'. This type of musical murmur is also found in acute myocardial infarction, syphilitic AI and rarely in acute rheumatic fever.

Can MI produce apical mid diastolic murmur ?

Yes. The increased flow through the valve in diastole may produce a low-pitched mid-diastolic rumbling murmur (functional MS), often with a S_4 .

Radiation of murmur if posterior mitral leaflet is affected :

In *posterior mitral leaflet* affection, pansystolic murmur of MI radiates to the anterior precordium and upto the pulmonary area. In a classical case of MI with radiation of murmur towards the left axilla and inferior angle of left scapula, the pathology remains in the *anterior mitral leaflet*. Rheumatic MI predominantly affects anterior mitral leaflet.

What is 'bruit'?

When the murmurs occur at the site of arterial stenosis, they are traditionally called bruits, e.g., renal artery bruit in renal artery stenosis. A short systolic bruit is produced in 50% or less obstruction, continuous bruit in 75% or more obstruction, and there is no bruit in 100% obstruction.

Differential diagnosis (D/D) of your case :

1. Functional murmur - Usually soft, not pansystolic, do not radiate to the axilla and without any thrill. No alteration with change of posture and respiration.
2. VSD - History starts from early childhood. Rough, pansystolic murmur, best audible over the lower left parasternal area; radiation to right chest. Very often associated with systolic thrill.
3. Conducted murmur of aortic stenosis - Harsh, mid-systolic murmur.
4. Tricuspid incompetence (TI) - Murmur best audible over the tricuspid area with positive Carvallo's sign (increase in intensity of murmur with inspiration), not conducted to the axilla, and may be associated with features of CCF (in functional TI, search for pulsatile liver and engorged-pulsatile neck vein).

Definition of pansystolic or holosystolic murmur :

They start immediately with the S and continue through to the S₂ (pansystolic). They always have a uniform intensity and thus known as holosystolic.

Why the murmur of MI is pansystolic ?

As soon as the LV pressure exceeds LA pressure, i.e., JUST AFTER THE S_j, the regurgitant jet of blood goes from LV to LA and CONTINUES TO AND THROUGH THE S₂ until the LA pressure exceeds LV pressure. This is why the murmur of MI is pansystolic in nature.

How to describe the 'intensity' or 'grading' of a murmur ?

The intensity of murmurs are divided into SIX GRADES (Levine's grading system). Grade I is a very faint murmur heard on careful auscultation, and grade VI murmur may be heard without a stethoscope and with the head just away from the chest (loudest).

If the murmur is associated with thrill, the grading is IV or IV+ (i.e., it may be grade IV, V or VI). Thrill is absent in grade I, II, and III. Diastolic murmurs are only graded upto IV.

Grade I — Very faint or soft (can be heard only with special effort)

Grade II — Soft

Grade III — Moderate

Grade IV — Loud with presence of thrill

Grade V — Very loud with associated thrill

Grade VI — Extremely loud and may be heard without a stethoscope; with associated thrill.

How to assess the 'severity' of MI clinically ?

Severe MI is assessed at the bedside by :

1. Presence of apical mid-diastolic flow murmur,
2. Left ventricular (apical) S₃,
3. Degree of LVH, and
4. Presence of systolic thrill with a loud murmur over the apical area.

Surface marking of mitral valve :

It is obliquely placed behind the left 4th costal cartilage and the adjacent part of the sternum.

What is mitral area ?

It corresponds with the apex of the heart. Normally in an adult, the apex is situated in the left 5th ICS, 1/2" inside the left MCL. Suppose, in a patient the apex beat is palpable in the left 6th ICS, 1/2" outside the MCL (in LVH) - this (apical) area will be the mitral area of the patient. So mitral area is a mobile area according to the position of the apex or apical area, i.e., it is not a fixed area. Tricuspid, aortic and pulmonary areas are always fixed in their location. See page 418 for other details.

When the patient is turned to left side while examining the CVS ?

1. To note the 'character' of apex beat,

2. For better palpation of the diastolic thrill (MS), and
3. For better auscultation of the diastolic murmur (MS).

* It is also said that in a case of MI, palpation of the systolic thrill and auscultation of the systolic murmur are better perceived in left lateral position. Never turn the patient to the left to localise the site of apex beat.

Palpate the mitral area :

Following four points are to be noted :

- a) Site of apex beat.
- b) Character of apex beat.
- c) Palpable heart sound (M₁).
- d) Any thrill.

Steps :

1. First locate the apex by proper clinical method described in page 21.
2. Then note the character of the apex beat — Turn the patient to left lateral position.
3. Next, one should confirm whether any palpable heart sound is present or not.
4. Lastly, feel for any thrill (one should put his left thumb over the right carotid) - if there is any difficulty, turn the patient to left lateral position and try to palpate the thrill in a better way at the height of expiration.

Complications of MI:

1. Acute LVF.
2. Infective endocarditis (more common than MS).
3. CCF.
4. Arrhythmias :
 - (i) Ventricular ectopics.
 - (ii) Atrial fibrillation (with embolic manifestations).
5. Giant left atrium may produce :
 - (i) Pressure symptoms — dysphagia, hoarseness of voice.
 - (ii) Atrial fibrillation.
6. Recurrent respiratory tract infections.

How do you like to investigate a case of MI ?

- (A) Chest X-ray (PA view) :
 1. LVH.
 2. LAH (giant left atrium).
 3. Signs of pulmonary venous hypertension (in acute MI).
 4. RVH.
- (B) ECG : The ECG may be normal in early stages. Following abnormalities may be seen :
 1. LAH (approximately in all chronic cases).
 2. LVH (50% cases).
 3. Biventricular hypertrophy (25% cases).
 4. Atrial fibrillation.
- (C) Curved barium-filled oesophagus pushed by enlarged LA in RAO view of chest X-ray.
- (D) Blood for (to detect active rheumatic carditis) :
 1. TC and DC 2. ESR 3. ASO titre
- (E) Echocardiography (giant or aneurysmal left atrium may be seen).
- (F) Doppler study (it quantifies the regurgitation).
- (G) Cardiac catheterisation with left ventriculography.

Treatment of MI:

1. Medical treatment — As in MS. MI of moderate severity can be managed medically. For the case described at the outset, digitalis and diuretic should be given to treat CCF; penicillin prophylaxis against SBE is a must. Digitalis and anticoagulants are added if AF is present. Vasodilators (nitrates, ACE-inhibitors) can reduce the regurgitant flow and may be helpful in severe cases.

2. Surgical treatment — Implantation of prosthesis (mitral valve replacement) is done under open heart surgery; sometimes, repair by mitral annuloplasty or valvuloplasty is done.

Features of digitalis toxicity :

1. Non-cardiac - Anorexia, nausea, vomiting (due to direct stimulation of CTZ at medulla), and diarrhoea; these are the earliest signs of toxicity.
2. Cardiac -

(i) Premature ventricular beats (bigeminy and multiform).	(iv) Ventricular tachycardia.
(ii) Varying degree of AV block (bradycardia).	(v) Bidirectional tachycardia.
(iii) Paroxysmal atrial tachycardia with block.	(vi) Sinus arrest.
	(vii) Ventricular fibrillation.
3. Chronic toxicity -

(i) Weight loss.	(iv) Xanthopsia (yellow vision).
(ii) Cachexia.	(v) Agitation, psychosis, delirium
(iii) Gynaecomastia.	

* Digitalis toxicity is managed by withholding digoxin. withdrawal of diuretic (hypokalaemia increases digitalis toxicity), giving potassium, administration of lignocaine or phenytoin to combat ventricular arrhythmias, and repeated application of digoxin antibody (Fab fragments).

What is cardiac output, cardiac index and ejection fraction ?

(A) CARDIAC OUTPUT = Stroke volume (output per ventricle per beat) X heart rate.

Normal - 5 - 6 litre/minute

(B) CARDIAC INDEX = Cardiac output / minute / square meter of body surface

Normal : 3.3 litre / minute / m²

End-diastolic volume - End-systolic volume

(C) EJECTION FRACTION ----- $\frac{\text{End-diastolic volume} - \text{End-systolic volume}}{\text{End-diastolic volume}}$ -----

Normal : 55-80%

Mild left ventricular dysfunction : 40-50%

Moderate left ventricular dysfunction : 30-40%

Severe left ventricular dysfunction : < 30%

Why history of drug intake is important in cardiology ?

1. Drugs can aggravate a disease ((β -blockers in heart block, thyroxine in angina, amlodipine in oedema, salbutamol in palpitation, α -blockers in postural hypotension, steroids in systemic hypertension).
2. Drugs can mask a disease (antihypertensives in systemic hypertension, diuretics in MS).
3. Drugs can modify a disease (oral contraceptive pills in systemic hypertension).
4. Drugs can cure a disease or alter physical sign (diuretics abolish the gallop rhythm in heart failure).
5. Drugs can induce a disease ((β -blockers in hyperlipidaemia, alcohol in cardiomyopathy).

N.B. : Diastolic blood pressure in MS is always very low. A diastolic blood pressure of 60 mmHg is common.

Case 3

AORTIC STENOSIS

What is your diagnosis ?

This is a case of valvular aortic stenosis (AS) of (AS) of atheromatous origin with congestive cardiac failure and the patient is in sinus rhythm at present.

What is your case ?

Build up the summary. The chief presenting symptoms are exertional dyspnoea, anginal pain, syncope and features related to LVF. All the symptoms are usually 'exertional'.

- a) Dyspnoea is primarily due to elevation of the pulmonary capillary pressure, pulmonary oedema; later on PND may occur. Severe AS may exist for many years without producing any clinical disability.

- b) Anginal pain is due to low aortic blood pressure and low cardiac output. Increased O_2 demand due to increased muscle mass (LVH) is also important. Thus, coronary blood flow cannot be maintained while on exertion.
- c) Syncope or black-out may be due to low and fixed cardiac output, LVF, low BP or transient atrioventricular block (all causing cerebral hypoperfusion). Sometimes, the patient complains of light-headedness, faintness or giddiness.

* *Dyspnoea, angina pectoris and syncope are three cardinal symptoms of AS; sometimes, a patient may be brought in the hospital emergency with 'sudden cardiac death'.*

Why do you say aortic stenosis ?

Because there is presence of :

(A) Symptoms (from the history) -

- a) Pain chest for 1 year.
- b) Exertional dyspnoea for 9 months.
- c) Occasional black-out for last 2 months.
(plus past H/O rheumatic fever).

(B) Signs -

- a) Pulse - 72 / min, low volume, regular, no radio-radial and radio-femoral delay, arterial wall is not thickened, all the peripheral pulses are palpable and of 'anacrotic' in type (also called pulsus parvus et tardus). 'Carotid shudder', i.e., a systolic thrill in the carotid artery is felt.
- b) BP - Low (say, 90/65 mm of Hg.) with low pulse pressure (in supine position).
- c) Apex beat - In normal position (i.e., left 5th ICS, 1/2" inside the left MCL) and heaving in character (though there is LVH, apex is usually not shifted because AS causes concentric hypertrophy). There is no palpable heart sound, no thrill in the apical area.
- d) Palpation of the aortic area - Systolic thrill felt. Best felt in full expiration with the patient sitting and leaning forward. The thrill is radiated to the carotid artery.
- e) Auscultation of the aortic area —
S₁ - Audible
S₂ (A₂) - Muffled
Ejection click — Present

A harsh midsystolic ejection murmur of grade V with direction of selective propagation towards the carotids is heard. The murmur is best audible in full expiration with the patient sitting and leaning forward, and with the diaphragm of stethoscope (occasionally the murmur of AS is transmitted to the mitral area and is known as *Gallavardin's phenomenon*).

- f) Auscultation of other areas of heart and lung—Within normal limit. No adventitious sound heard.

(g) Examination of respiratory system, G. I. tract and nervous system revealed no abnormality).

* The murmur of AS is known as saw-like murmur or crescendo-decrescendo murmur (never say diamond shaped' murmur as this is the phonocardiographic pattern of AS murmur).

** Add features of pulmonary hypertension, if present.

*** The normal aortic valve area is 2.5-3.5 cm². If it is < 1 cm² or having a gradient > 50 mm of Hg (normally, no gradient), the stenosis is critical.

****The pulse is not only small in volume but is slow in rising to a peak (plateau pulse).

Why the murmur of AS is midsystolic ?

The pressure gradient between the left ventricle and aorta is greatest in the middle of systole, and this is why the murmur of AS is midsystolic.

Significance of ejection click :

Ejection click is a sharp and high-pitched clicking sound heard immediately after the first heart sound due to sudden opening of the aortic or pulmonary valve (semilunar valves), and are best audible in aortic (in aortic stenosis) and pulmonary area (in pulmonary stenosis) respectively. **Its presence indicates that the stenosis is at the 'valvular' level and the stenosis (aortic stenosis or pulmonary stenosis) is of milder degree.** Pulmonary ejection click in pulmonary stenosis becomes less audible (soft) in inspiration, in contrast to all other right-sided events.

Example of 'non-ejection click' is mitral valve prolapse syndrome (MVPS).

Aetiology of AS :

1. **Rheumatic** (it should be remembered that isolated rheumatic AS is rare: usually it is associated with MS, AI or MI — So, try to find them out. Aortic valve is the second most frequently affected valve in rheumatic fever after mitral valve).
 2. Congenital (commonly bicuspid aortic valve) - Young adult patients.
 3. Calcific degeneration of the aortic valve (old age).
 4. Associated with familial hypercholesterolaemia, mucopolysaccharidosis.
 5. Functional — In severe AI, thyrotoxicosis, severe anaemia etc.
- * Calcific valvular disease is the commonest cause of AS.

Types of aortic stenosis :

- (A) Valvular — Examples are described above in 'Aetiology of AS'.
 - (B) Supravalvular — May be associated with Elfin facies, mental retardation, hypercalcaemia.
 - (C) Subvalvular (special variety is IHSS) — Idiopathic hypertrophic subaortic stenosis.
- * Elfin facies — Broad forehead, pointed chin, cupid's bow-like upper lip, upturned nose, hypertelorism and low set ears (seen in supravalvular AS and when associated with mental retardation and hypercalcaemia, it is known as William's syndrome).

Causes of RVH :

1. Pulmonary hypertension (specially MS and other left-sided valvular diseases, COPD).
2. Pulmonary stenosis.
3. Cardiomyopathy.
4. ASD, VSD.
5. Fallot's tetralogy.
6. Hyperdynamic circulation.

What are the types of LVH ?

(A) **CONCENTRIC** (muscle mass increases at the cost of left ventricular cavity i.e., left ventricle is more hypertrophied than dilated - so the apex is not shifted much). The examples are basically left ventricular outflow tract obstruction, and are :

- | | |
|----------|---------------------------|
| 1. AS. | 3. Coarctation of aorta. |
| 2. IHSS. | 4. Systemic hypertension. |

(B) **ECCENTRIC** (left ventricular dilatation as well as increase in its muscle mass - so the apex goes down and outwards). The examples are :

- | | |
|---|------------------------------|
| 1. Systemic hypertension. | 5. Thyrotoxicosis. |
| 2. MI or AI. | 6. Cardiomyopathy (dilated). |
| 3. Ischaemic heart disease (IHD). | 7. VSD, PDA. |
| 4. Severe anaemia (hyperdynamic circulation). | |

* **Combined A + B are the causes of LVH.**

** Hypertrophy of ventricle is due to pressure overload (e.g., systemic hypertension) and dilatation is the result of volume overload (e.g., AI, MI).

Causes of right atrial hypertrophy :

1. Tricuspid stenosis.
2. Tricuspid incompetence.
3. RVH of some duration.

Causes of left atrial hypertrophy :

1. MS.
2. MI (giant left atrium may be seen).
3. LVH of some duration.

Causes of biventricular hypertrophy (LVH + RVH) :

1. Cardiomyopathy (dilated).
2. Ventricular septal defect.

3. Myocarditis.
4. Ischaemic heart disease.
5. Systemic as well as pulmonary hypertension.
6. Hyperkinetic circulation (e.g., in severe anaemia).

Commonest cause of displaced apex :

Deformity of thoracic cage, usually scoliosis. So, never forget to inspect the back in any CVS case.

Thrill in different areas of the heart :

(A) MITRAL AREA :

- a) Systolic -
 1. MI (commonest).
 2. VSD.
 3. ASD (ostium primum type).
- b) Diastolic -
 1. MS (commonest).
 2. Left atrial myxoma (very rare).

(B) AORTIC AREA :

Congenital or acquired AS (thrill in aortic area is almost exclusively systolic). Rarely, there is diastolic thrill of AI.

(C) PULMONARY AREA : Usually it is systolic thrill—

1. Pulmonary stenosis.
2. ASD.
3. High VSD.
4. Fallot's tetralogy.
5. PDA (sometimes, continuous thrill).

* Graham Steell murmur is usually not associated with diastolic thrill.

(D) TRICUSPID AREA : Usually it is systolic thrill—

1. VSD.
2. Tricuspid incompetence.
3. ASD (ostium primum).
4. Pulmonary stenosis (infundibular type).

Causes of systolic murmur in base of the heart :

(A) AORTIC AREA :

1. Aortic stenosis (AS).
2. Coarctation of aorta.
3. Systemic hypertension.
4. Functional (from AI).
5. Aneurysm of aorta.
6. Aortic sclerosis (old age).

(B) PULMONARY AREA :

1. Pulmonary stenosis (PS).
2. Pulmonary hypertension.
3. Hyperkinetic circulatory states
4. ASD.
5. Fallot's tetralogy.
6. Innocent murmur.

Classify 'organic' murmurs :

(A) SYSTOLIC :

1. Ejection systolic — Starts after the S₁ and ends before the S₂. Examples are AS, PS and ASD.
2. Pansystolic — Starts immediately with the S₁ and continues through to the S₂, and typically they are of uniform intensity (holosystolic), e.g., MI, TI and VSD.
3. Late systolic, e.g., MVPS, IHSS, papillary muscle dysfunction.

(B) DIASTOLIC :

1. Early diastolic — AI, and PI (Graham Steell murmur).
2. Mid-diastolic — MS, TS, Austin Flint murmur, Carey-Coombs murmur, and flow murmur in ASD or VSD.
3. Presystolic or late diastolic — MS, atrial myxoma.



Palpation of base of the heart (in sitting position)



Palpation of trachea



Percussion of the left apex of lung from behind



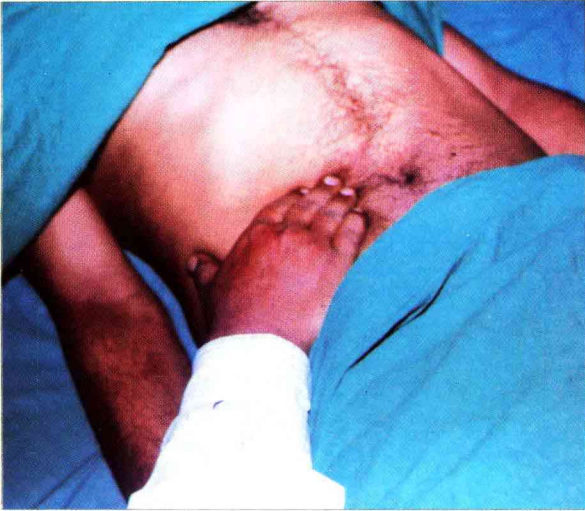
Examining movement of the chest from back (apical area)



Examining movement of the chest (lower anterior part)



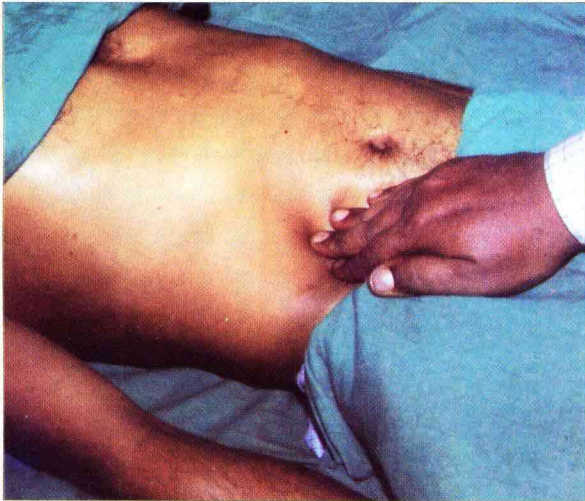
Examining movement of the chest (upper anterior part)



Palpation of liver (conventional method)



Palpation of liver (preferred method)



Palpation of liver (alternative method)



Palpation of spleen



Palpation of spleen (just palpable) in right lateral position



Palpation of spleen (just palpable) by hooking from left side

(C) CONTINUOUS :

1. PDA.
2. Rupture of sinus of Valsalva.
3. Pulmonary arteriovenous fistula.
4. Aortopulmonary window.
5. Coronary arteriovenous fistula.

Innocent murmur :

These are commonly found in **children** (Still's murmur) and usually disappear as the child grows older. Here, the heart with its valves are absolutely normal. Possible mechanisms are :

- a) Hyperkinetic circulatory states in children due to increased heart rate, and
- b) Increased resistance of the pulmonary vascular bed in children.

The characteristics are :

1. Usually systolic; soft (upto grade II); localised.
2. No thrill.
3. Best heard over the pulmonary area (mainly along upper left sternal border).
4. Heard best in supine position and may disappear in upright position; often the murmur is heard after exercise, crying or fever.
5. Associated with normal heart sounds, chest X-ray and ECG.

Haemic murmur :

Sometimes, an ejection systolic murmur is heard in pulmonary area when the person suffers from severe anaemia (so called haemic). The possible mechanisms are :

- a) Increased flow of blood through the pulmonary artery, and
- b) Dilatation of the pulmonary artery.

These mechanisms also play their part in other hyperkinetic circulatory states like pregnancy, thyrotoxicosis, high fever etc. Often severe anaemia produces mitral systolic murmur due to ring (mitral) dilatation. This type of murmur disappears after correction of the primary disease. They are never associated with abnormality in ECG. Sometimes, a systolic thrill is associated with haemic murmur.

Functional murmur :

In functional murmur, there is no organic heart disease present at the site of production of murmur and is produced only due to increased flow of blood. Many clinicians know these murmurs as 'flow murmurs'.

The common examples are :

1. Systolic murmur in the pulmonary area in a case of ASD.
2. Early diastolic Graham Steell murmur of pulmonary hypertension (functional pulmonary incompetence).
3. Functional systolic murmur in aortic area (functional AS) in a case of severe AI.
4. Apical mid-diastolic murmur in a case of severe AI (Austin Flint murmur).

Characteristics :

1. Usually systolic, may be diastolic.
2. Intensity of murmur — Not very loud.
3. Commonly without a thrill (remember, presence of thrill does not exclude functional murmur).
4. Not distributed to a wide area (more or less localised).
5. Does not change much with change of posture.
6. Does not produce cardiomegaly (haemodynamically insignificant).
7. Murmur disappears after correction of the increased flow.

Changing murmur :

These are the murmurs which change their character or intensity from time to time, e.g.,

1. Infective endocarditis,
2. Atrial myxomas, and
3. Atrial thrombus.

How to assess the 'severity' of AS clinically ?

Severe AS is assessed at the bedside by the presence of :

1. H/O effort dyspnoea, effort syncope, effort angina.
2. Low volume pulse with a slowly rising plateau (e.g., delayed carotid upstroke).
3. Systolic decapitation of BP (low systolic pressure due to low and fixed cardiac output; in a patient with isolated AS, systolic BP never goes above 150 mm of Hg.).
4. Paradoxical splitting of S_2 — the splitting happens to occur only during expiration.
5. Presence of S_4 .
6. Absence of systolic thrill often tells against a severe stenosis.
7. Harsh, loud and long murmur with late systolic peaking.

Splitting of S_2 ?

It is of three types —

1. Physiological—When the splitting is present only during inspiration, it is physiological splitting. Here A_2 occurs earlier than P_2 .
2. Reversed (paradoxical)—Splitting occurs only during expiration.
3. Pathological—Splitting of S_2 is audible both in inspiration and expiration.

Causes of wide and fixed (no change with respiration) splitting of S_2 :

1. Atrial septal defect (hallmark of ASD),
2. CCF (severe right heart failure),
3. Massive pulmonary thromboembolism,
4. Total anomalous pulmonary venous connection (TAPVC),
5. Right ventricular pacing.

Causes of wide but variable splitting of S_2 :

1. RBBB (right bundle branch block).
2. Pulmonary stenosis.
3. Left ventricular pacing.
4. Ventricular septal defect (VSD).

Reversed (paradoxical) splitting of S_2 :

1. Severe AS.
2. LBBB (left bundle branch block).
3. Systemic hypertension.
4. Severe left ventricular dysfunction.

Single S_2 :

1. Severe AS (with absent A_2), aortic atresia (absence of aortic valve).
2. Severe PS (with absent P_2), pulmonary atresia (absence of pulmonary valve).
3. Fallot's tetralogy.

Causes of loud S_2 :

- (A) Loud P_2 — Pulmonary hypertension, pulmonary artery dilatation.
- (B) Loud — Systemic hypertension, dilated aorta, aneurysm of aorta, hyperkinetic circulatory states.

t Palpable S_2 (either P_2 or A_2) is known as **diastolic shock** or **diastolic knock**.

** Soft P_2 , and A_2 occur respectively due to PS, and AS.

*** Loud (both) P_2 and A_2 may be found in patient with thin chest wall, and combined pulmonary hypertension plus systemic hypertension.

**** Normally the S_2 is short and high-pitched (dup in 'lubb-dup'), and is best audible in the pulmonary (P_2) and aortic area (A_2). S_2 indicates the beginning of ventricular diastole and is produced due to closure of aortic (A_2) and pulmonary (P_2) valves. In health, there is a gap of 0.03 second between A_2 and P_2 . Normal physiological splitting of S_2 is audible in children and young adults in inspiration.

Murmur of bicuspid aortic valve :

Usually the AS-murmur is heard but sometimes, AI-murmur is also present. The AI-murmur is haemodynamically insignificant and in bicuspid aortic valve, features of AS is always greater than AI.

Who invented the stethoscope ?

The French physician R.T.H. Laennec (1816).

Bedside diagnosis of 'acute rheumatic carditis' :

This is a '**PANCARDITIS**' involving endocardium, myocardium and pericardium to varying degree. The features are :

(A) Pericarditis :

1. Pericardial rub.
2. Pericardial effusion (rare and always mild).

(B) Myocarditis :

1. Relative tachycardia (pulse rate rises by more than 10/minute with per degree (°F) rise of temperature).
2. Tic-tac' quality of heart sounds (due to tachycardia and mimic foetal heart sounds).
3. S₁ — Muffled or soft (reflected as prolonged PR interval in ECG).
4. S₃ gallop rhythm due to heart failure.
5. Congestive heart failure.
6. Heart size — Enlarged (apex goes down and outwards due to cardiac dilatation). It is a very important clinical sign.
7. Conduction defects — Dropped beat in pulse due to heart block.
8. Arrhythmia — May occur.

(C) Endocarditis :

1. The appearance of —
 - a) Soft systolic murmur due to mitral incompetence.
 - b) Soft early diastolic murmur in the aortic area due to acute involvement of the aortic valve (murmur of AI).
 - c) Carey-Coombs murmur — Due to mitral valvulitis (temporarily producing MS as a result of oedema of the mitral valve cusps),

or, 2. Change in the character of existing organic heart murmurs.

* Rheumatic subcutaneous nodules are almost always associated with active carditis.

** An accelerated heart beat during rest or sleep make the clinician suspicious of carditis.

Appearance of murmur in rheumatic valvulitis (carditis) from acute rheumatic fever :

In order of sequence :

1. MI murmur.
2. AI murmur.
3. Carey-Coombs murmur.

Diagnosis of acute rheumatic fever :

It is done by revised **JONES CRITERIA**, updated in 1992 (after Dr. T. Duckett Jones) —

(A) MAJOR MANIFESTATIONS :

1. Carditis
2. Polyarthritides
3. Chorea
4. Erythema marginatum
5. Subcutaneous nodules

(B) MINOR MANIFESTATIONS :

1. Fever
2. Arthralgia
3. Previous rheumatic fever
4. High ESR or positive C-reactive protein (CRP)
5. Leucocytosis
6. Increased PR interval on ECG

The presence of,

- a) Two or more major, or
- b) One major + 2 or more minor criteria strongly suggest rheumatic fever

WITH

One supporting evidence of preceding streptococcal infection such as :

- a) H/O recent scarlet fever,
- b) Positive throat culture for group A streptococcus,
- c) Increased ASO (antistreptolysin O) titre or titre of other streptococcal antibodies.

* Do not forget to take the H/O sore throat, 2-4 weeks prior to the onset of acute rheumatic fever. Evidence of recent streptococcal infection is particularly valuable if only one major manifestation is present.

** Acute rheumatic fever develops as a delayed sequel to pharyngeal infection caused by group A beta-haemolytic streptococci (type 5 most commonly causes rheumatic fever; other rheumatogenic serotypes are 1, 3, 6, 14, 18, 19 and 24). Streptococcal skin infection never produces rheumatic fever.

*** The WHO (2002-2003) criteria for the diagnosis of rheumatic fever is a minor variation of Jones criteria (1992).

Investigations done in acute rheumatic fever :

- (A) Blood ;
 1. Anaemia, polymorphonuclear leucocytosis, raised ESR.
 2. Raised ASO titre (> 200 units in adult and > 300 units in children).
 3. Raised C-reactive protein and other acute phase reactants (e.g., serum complements).
 4. Other streptococcal antibodies — Antihyaluronidase (AH), anti-streptokinase (ASK), anti-streptozyme (ASTZ), and anti-DNase B (antideoxyribonucleotidase B). Now-a-days, anti-DNase B and ASO titre are preferred for diagnosis.
 5. Positive throat swabs culture for Group A streptococci in 25-40% cases.
- (B) ECG (evidence of carditis) :

1. Sinus tachycardia.	4. T wave inversion.
2. Prolonged PR interval.	5. Reduction in QRS voltage.
3. Features of pericarditis.	
- (C) Chest X-ray :
Cardiomegaly, pulmonary congestion or evidences of heart failure.
- (D) Echocardiography : Cardiac dilatation with valve abnormalities (in case of carditis).

Latent period from rheumatic fever to organic valvular heart disease :

In temperate climates, this gap is usually 15 years. In India, organic disease (mainly MS) develops much earlier (latent period may be as short as 1-2 years), and frequently causes serious symptoms before the age of 20 years. This happens to be due to repeated attacks of severe carditis in India.

Characteristics of rheumatic arthritis :

1. Most common in children (5 to 15 years); commonest 'major' manifestation (75%).
2. Big joints are involved (knee, most commonly); but no joint is immune to the inflammatory process. Small joints of hand and feet may be affected as seen in rheumatoid arthritis. Joint involvement is often symmetrical. Involved joints are swollen, painful and tender.
3. Typically it is fleeting or migratory in nature (as the inflammation of one joint is subsided, others tend to become affected); the affection of joints in rheumatoid arthritis is 'additive in nature'.
4. Spine, sternoclavicular and temporo-mandibular joints are rarely affected (affected more in rheumatoid arthritis).
5. Recovery is complete.
6. Usually there is no residual deformity (rarely, recurrent attacks of arthritis may lead to minor deformities of the hand at MCP joints — Jaccoud's arthritis). Deformity is very common in rheumatoid arthritis.
7. Muscle spasm and muscular wasting are not common (common in rheumatoid arthritis).
8. No radiological abnormality (rheumatoid arthritis have characteristic X-ray features).

* **Arthritis** means inflammation of the joint, i.e., pain plus swelling of the joint. **Arthralgia** means subjective presence of pain only (no inflammation, no swelling). *Acute rheumatic fever bites the heart and licks the joint in children, whereas it bites the joints and licks the heart in adults.*

Possible causes of fleeting or migratory arthritis :

1. Rheumatic arthritis (commonest).
2. SLE.
3. Drug reaction/serum sickness.
4. Arthritis following gonococcal or meningococcal bacteraemia.
5. Viral arthritis (Lyme arthritis, Chikungunya).
6. Arthritis associated with inflammatory bowel disease or Whipple's disease.
7. 'Seroconversion' phase in AIDS.
8. Septicaemia.
9. Following intestinal by-pass surgery.
10. Sarcoidosis.

Active rheumatic carditis with low ESR—reason behind :

The patient is in heart failure.

Causes of very low ESR :

- | | |
|---|--------------------------------------|
| 1. Polycythemia (COPD, Fallot's tetralogy). | 3. Congenital fibrinogen deficiency. |
| 2. Congestive cardiac failure. | 4. Sickle cell anaemia. |

Causes of high ESR :

- | | |
|---|-----------------------------------|
| 1. All acute infections (e.g., tuberculosis). | 6. Malignancy. |
| 2. Severe anaemia. | 7. Multiple myeloma (stormy ESR). |
| 3. Pregnancy. | 8. Vasculitis. |
| 4. Old age. | 9. Acute rheumatic carditis. |
| 5. Collagen vascular diseases (SLE). | 10. Temporal arteritis. |

* Very high ESR is seen in multiple myeloma, temporal arteritis and advanced tuberculosis.

** Normal ESR in men is 0-15 mm/hr and in women it is 0-20 mm/hr. ESR reflects changes in plasma proteins. Some clinicians use ESR as 'sickness index.'

What are acute phase reactors ?

These are a class of liver-produced proteins, which rise in response to cytokines synthesized in tissue injury e.g., in inflammation. Phase reactors are fibrinogen, haptoglobin, ferritin, α_1 -antitrypsin, serum amyloid-A, mannose-binding protein, C_3 complement, ceruloplasmin, C-reactive protein and orosomucoid. In clinical practice, estimation of C-reactive protein is commonly done. They are responsible for rise in the ESR. Phase reactors serve as markers for 'disease activity'.

Differential diagnosis (D/D) of your case :

It is a patient of valvular AS (due to the presence of ejection click and soft A_2). The D/D are :

1. Subvalvular aortic stenosis—Characterised by,
 - a) Early diastolic murmur of AI (common),
 - b) No ejection click,
 - c) In IHSS variety - double kicking apex beat and pulsus bisferiens may be present,
 - d) The murmur does not radiate to carotids,
 - e) No post stenotic dilatation,
 - f) Heart size tends to be larger.
2. Supravalvular aortic stenosis—Characterised by,
 - a) Systolic BP, of right arm > left arm,
 - b) No ejection click,
 - c) Thrill transmitted to carotids (thrill is more conspicuous than other types),
 - d) May be associated with mental retardation, Elfin facies, squint, infantile hypercalcaemia,
 - e) A_2 - Normal / accentuated,
 - f) No post stenotic dilatation.

* Also consider *other causes of systolic murmur* in the aortic area (page 30 and 31).

Complications of AS :

1. Acute LVF.
2. Right-sided heart failure (CCF).

3. Infective endocarditis.
 4. Tachy-and bradyarrhythmias.
 5. Precipitation of angina pectoris or syncope.
 6. Sudden death due to complete heart block or ventricular fibrillation.
- * Bleeding per rectum resulting from angiodysplasia of colon may be associated with AS.

How do you like to investigate a case of AS ?

1. X-ray chest (PA view) :
 - a) Normal sized heart due to concentric hypertrophy of left ventricle or LVH.
 - b) Post stenotic dilatation of ascending aorta.
 - c) Calcification of aortic valve (in left anterior oblique view or on fluoroscopy).
2. Fluoroscopy : Aortic valve calcification is seen.
3. ECG :
 - a) LVH.
 - b) LAH.
 - c) First degree heart block.
 - d) LBBB, complete heart block (when calcification extends into conducting system).
4. Blood for (to detect active rheumatic carditis) :
 - a) TC and DC b) ESR c) ASO titre
5. Echocardiography shows calcified aortic valve, LVH; doppler estimate of gradient etc.
6. Cardiac catheterisation : to identify associated coronary artery disease.
7. Coronary angiogram : To rule out IHD in patients of AS with angina pectoris.

How will you like to treat this case ?

- By, 1. Medical management — as in MS. Treatment of angina, atrial fibrillation and LVH should be done carefully. | 3-blockers should preferably be avoided.
2. Asymptomatic AS — Review regularly for development of angina, syncope or heart failure.
 3. In childhood or adolescence — valvotomy; adults and elderly—valvuloplasty.
 4. For calcific AS — Aortic valve replacement (prosthesis).

Describe the classical pain of angina pectoris :

Characteristics :

1. Commonly retrosternal; sometimes, felt in left side of the midline.
2. Constricting, pressing or squeezing in nature; along with a sense of choking. Often described as 'like a band around the chest'.
3. Radiation — Both upper extremities (left > right), shoulders, lower jaw, neck, back, rarely to the upper abdomen. Radiation to the ulnar aspect of the left arm is characteristic.
4. Exertional commonly. Often increases with emotion, exposure to cold or walking uphill after a heavy meal.
5. Duration is usually 2-5 minutes; seldom lasts more than 20 minutes.
6. Relieved by rest, or after taking sublingual nitrates or nitroglycerin.
7. Usually associated with palpitation and diaphoresis.

N.B. ; A chest pain present in the left inframammary region is usually not due to IHD. The pain of myocardial infarction is more severe, lasts longer, persists at rest, and very often associated with nausea, vomiting, diaphoresis and pallor. If the anginal pain occurs at rest ('rest pain') think of acute myocardial infarction, unstable angina, vasospastic angina (Prinzmetal's angina) and emotion-induced angina.

The diagnosis of ischaemic chest pain depends on history and different investigations (ECG, stress testing, thallium stress test, Holter monitoring, echocardiography and Doppler study, and the most specific coronary angiography).

Syncope, fainting or black-out — definition and aetiology :

Definition - Syncope is sudden and transient loss of consciousness with inability to stand upright as a result of impairment of cerebral blood flow (i.e., postural collapse).

Aetiology -

1. Simple syncope, i.e., due to vasovagal phenomenon (in an unpleasant sight, micturition syncope, cough syncope, carotid sinus syncope or acute severe pain due to any cause).
2. Cardiovascular (complete heart block, AS, IHSS, sick sinus syndrome, hypersensitive carotid sinus syncope, prolonged QT syndromes, acute pulmonary thromboembolism, Eisenmenger's syndrome, other brady- or tachyarrhythmias, left atrial ball valve thrombus or myxoma).
3. Metabolic or drug-induced (hypoglycaemia, hypocalcaemia, alcohol intoxication; methyl dopa, prazosin, vasodilators or diuretics-induced postural hypotension).
4. Postural hypotension (in patients receiving antihypertensives and vasodilator drugs).
5. Nervous system disorders (vertebro-basilar insufficiency, diabetic neuropathy, G.B.syndrome, familial dysautonomia, glossopharyngeal neuralgia).
6. Hyperventilation and hysterical fainting, breath-holding (children), unknown causes.

* Exertional syncope is classically seen in aortic stenosis.

D/D of syncope with episodic weakness/fainting :

Hyperventilation syndrome, hypoglycaemia, acute blood loss (hypovolaemia), vertigo, transient ischaemic attack, severe anaemia etc.

Describe the 'black-out' in this patient :

1. Present for last 2 months,
2. Persists for few seconds,
3. Almost always exertional,
4. No H/O trauma.

How to take the H/O dyspnoea in a patient ?**Following points should be noted carefully :**

1. Paroxysmal or exertional ? How much exertion is needed?
2. Any H/O PND ? or orthopnoea?
3. Seasonal variation present or not?
4. Associated with wheeze or not?
5. Grading of dyspnoea (NYHA classification).
6. Whether associated with cough or pain chest (pneumothorax) or shock (myocardial infarction), fever (consolidation), angina (AMI).
7. Time of appearance : early night (cardiac asthma), early morning (bronchial asthma).
8. Progressive or not?
9. How relieved? by drugs/rest/change of smoky environment/squatting / change of posture or by expectoration of tenacious sputum.

Enumerate the different causes of dyspnoea :

The major causes of dyspnoea are due to cardio-respiratory abnormalities ;

1. Respiratory : Bronchial asthma, COPD, airways obstruction, lobar and bronchopneumonia, spontaneous pneumothorax, pleural effusion, pulmonary thromboembolism, pulmonary oedema, bronchogenic carcinoma, fibrosing alveolitis, pulmonary infections (pneumonias).
2. Cardiovascular : Acute left ventricular failure (e.g., AMI, valvular heart disease), SVC syndrome, cyanotic congenital heart diseases, pericardial effusion and cardiac tamponade, constrictive pericarditis, rapid cardiac arrhythmias, IHD, cardiomyopathy.
3. Metabolic : Diabetic ketoacidosis, uraemia, high fever.
4. Neurological ; Depression of respiratory centre from motor neurone disease, GB syndrome, bulbar poliomyelitis, syringobulbia; raised intracranial tension; diaphragmatic or respiratory muscle paralysis, myasthenia gravis.
5. Mechanical : Huge ascites.

6. Psychogenic : Hysterical hyperventilation, anxiety neurosis.
7. Physiological : High altitude, exercise, severe anaemia.

Differentiation between dyspnoea of pulmonary and cardiac origin :

Dyspnoea due to pulmonary aetiology : Dyspnoea due to cardiac aetiology :

- | | |
|--|--|
| 1 Cough with expectoration precedes dyspnoea | 1. PND and orthopnoea |
| 2. Wheeze or stridor | 2. Dyspnoea precedes cough with expectoration |
| 3. Pleuritic chest pain | 3. Associated angina, syncope, palpitation; hypertension |
| 4. Pyrexia | 4. Rapid progression of symptoms |
| 5. Seasonal variation | 5. Little or no cyanosis (+) |
| 6. Cyanosis (++ or +++) | 6. Erect posture or squatting may relieve |
| 7. Progression over many years | 7. Oedema is an important sign |
| 8. Response to oxygen, bronchodilators | 8. Response to diuretics and digoxin |
| 9. Absence of obvious cardiac disease | 9. Physical signs of cardiac disease |

Management of acute rheumatic fever in a nutshell :

1. Bed rest - specially for fever, arthritis, arthralgia, carditis and heart failure.
2. High calorie, salt restricted diet.
3. Chorea - reassurance, sedatives like clonazepam or chlorpromazine, and in severe cases haloperidol, sodium valproate or carbamazepine is used.
4. Symptomatic treatment :
 - a) Arthritis, arthralgia, fever—aspirin in a dose of 80-100 mg/kg/day in children and 4-8 g/day in adults in 4-6 divided doses is started, and continued for 2 weeks. If symptoms subside, a lower dose of 60-70 mg/kg/day (in children) is continued for a further 2-4 weeks.
 - b) Patient without carditis — aspirin is preferred.
 - c) Patient with carditis but without heart failure — aspirin ± glucocorticoid (role of corticosteroids are doubtful in carditis but majority of cardiologists believe that they help in rapid resolution of heart failure). Prednisolone 1-2 mg/kg/day (maximum of 80 mg) is given orally for a period of 2 weeks and then tapered off over next 2 weeks.
 - d) patient with carditis and heart failure—majority opine that corticosteroid is mandatory over and above aspirin.
 - e) Antibiotics — single injection of 1.2 million units of benzathine penicillin, I.M is given to eradicate group A streptococcal infection, if present. Oral penicillin (penicillin V) 500 mg twice daily, orally for 10 days or erythromycin 40 mg/kg/day, orally may be used for 10 days (in patient allergic to penicillin).

5. Secondary prevention of acute rheumatic fever :

The mainstay of controlling rheumatic heart disease is secondary prevention. The dose of inj. benzathine penicillin is 1.2 million units, I.M given at 4 weekly interval (in endemic areas and high-risk cases, the interval is 2-3 weekly); dose of penicillin in children is 0.6 million units. Instead oral penicillin V may be used 250 mg twice daily or oral erythromycin 250 mg twice daily (in penicillin allergy). Controversies exist regarding duration of therapy and the guideline is—

- a) Rheumatic fever without proven carditis - 5 years after the last attack or until the age of 18 years, whichever is longer.
- b) Rheumatic fever with carditis but having minimal residual valve damage—10 years after the last attack or 25 years of age, whichever is longer.
- c) Rheumatic fever with carditis along with residual heart disease—10 years after the last episode or until the age of 40, whichever is longer. Few clinicians prefer to give penicillin life long in this situation.
- d) If valvular surgery is done—Penicillin prophylaxis is continued life long.

* Prophylaxis against infective endocarditis is also needed in valve damage.

Case 4

AORTIC INCOMPETENCE

What is your diagnosis ?

This is a case of valvular aortic incompetence (regurgitation) of rheumatic origin without congestive cardiac failure and the patient is in sinus rhythm at present.

What is your case ?

Try to build up the summary. Aortic incompetence (AI) remains asymptomatic for years or decades. Main symptoms of aortic incompetence are,

1. Palpitation,
2. Awareness of the heart beat, specially on lying; pulsatile or throbbing sensation.
3. Chest pain (angina pectoris, or due to pounding of heart against chest wall),
4. Dyspnoea (exertional),
5. Features of LVF or CCF (orthopnoea, PND, oedema etc).

Peripheral signs of AI (chronic) :

Start from hands and go to the neck, head and then downwards to the legs systematically in search of peripheral signs. **Peripheral signs are produced as a result of 'wide pulse pressure'**. They are :

1. Visible capillary pulsation (Quincke's sign) at nail-beds or lip i.e., blanching and quick refill—
 - a) When light pressure is applied to the tip of fingers or nails, there is alternate flushing and pallor of the nail-bed, or
 - b) When a glass slide is pressed on the everted lower lip, it produces alternate redness and blanching.
2. Prominent digital artery pulsation — Hold the flexed fingers of the patient with your fingers in flexed position to feel the pulsation.
3. High volume collapsing pulse or Corrigan's pulse (water-hammer pulse).
4. Locomotor brachialis — Look at the medial side of lower arm (with semiflexed elbow) for 'tortuous and highly pulsatile' brachial artery. Tortuosity is the most important component of locomotor brachialis and thus, it is commonly found in atherosclerosis (old age).
5. Blood pressure — Wide pulse pressure with low diastolic pressure (even Korotkoff sounds may continue upto 0 mm of Hg.). Pulse pressure is usually > 60 mm of Hg.
6. Corrigan's sign — Dancing carotids in neck.
7. de Musset's sign (named after a French poet) — To and fro head-nodding synchronous with carotid pulsation (systolic extension of neck).
8. Visible pulsation in the suprasternal (jugular) notch.
9. 'Pistol shot' sound or Traube's sign—Booming sound produced after pressing the stethoscope over femoral artery.
10. Systolic murmur on compression of the femoral artery proximally.
11. Duroziez's murmur — Diastolic murmur on compression of the femoral artery distally.
12. Hill's sign — Increase in the femoral artery systolic BP > 20 mm of Hg above the brachial artery systolic BP. In health, the systolic difference between two extremities remains within 20 mm of Hg. In severe AI, the increase is > 60 mm of Hg. It is very important and specific sign of AI.

* Pulsations in uvula, liver and enlarged spleen are respectively known as Muller's sign, Rosenbach's sign and Gerhardt's sign. Changes in pupil size with each cardiac systole is known as Landolfi's sign. Alternate flushing and blanching of forehead is known as Lighthouse sign, and pulsation of retinal artery is Becker's sign. Duroziez' sign is the combination of No. 10) and 11).

Why do you say aortic incompetence ?

Because there is presence of :

- (A) **Symptoms (from the history)** — Palpitation, throbbing sensation all over the body, chest pain, dyspnoea for last 2 years. Past H/O rheumatic fever is also present.

(B) **Signs —**

- a) Pulse — 80 / min, regular, high volume, neither there is any inequality nor any delay, arterial wall is not thickened, all the peripheral pulses are 'nicely' palpable and it is water-hammer in type.

- b) BP — Say, 145/40 mm of Hg. (in supine position), i.e., high systolic and low diastolic BP. So, there is wide pulse pressure.
- c) **All the peripheral signs are present** (described above).
- d) Apex — Diffuse, goes down and outwards (LVH) with hyperdynamic character.
- e) Palpation of the aortic area — No thrill, no palpable heart sound, no pulsation [though very rare, one may get a diastolic thrill (5%) in aortic area or in lower left sternal border].
- f) Auscultation of the aortic area—
S₁ — Audible
S₂ (A₂) — Muffled
Murmur— High-pitched, soft blowing, early diastolic decrescendo murmur of grade III which is best audible in sitting and leaning forward position, in full expiration and with the diaphragm of stethoscope. The murmur is radiated towards the apex and is best heard at neoaortic area (left 3rd ICS).
- * Occasionally, the murmur may radiate upto apex and left axilla, and is known as **Cole-Cecil murmur**. Sometimes, S₃ may be heard.
- g) Other areas of heart and lungs— Within normal limit.
- * Add features of pulmonary hypertension, if present. Always examine the neck for thyroid enlargement in a patient with wide pulse pressure.
- [h) Examination of respiratory system, G. I. tract and nervous system revealed no abnormality],
- * Patients of AI remains asymptomatic for years after years, but once decompensated, they start deteriorating rapidly.

Different types of murmur found in AI:

1. Classical early diastolic, soft blowing, high-pitched murmur in the aortic or neoaortic area.
2. Ejection systolic murmur of functional AS in the aortic or neoaortic area.
3. Austin Flint murmur — Mid-diastolic rumbling murmur (flow murmur) in the mitral area.

How to assess the 'severity' of AI ?

Bedside assessment of severe AI is done by,

1. Presence of marked peripheral signs.
2. Hill's sign of more than 60 mm of Hg (difference in systolic BP between lower and upper extremity 20-40 mm of Hg—mild AI, 40-60 mm of Hg—moderate AI, and > 60 mm of Hg— severe AI). Hill's sign is due to the phenomenon of recruitment of impulses.
3. Long duration of early diastolic murmur.
4. Presence of Austin Flint murmur.
5. Pulsus bisferiens (read the section on 'Water-hammer pulse').

Aetiology of AI:

- | | |
|----------------------------|---|
| 1. Rheumatic (80%). | 7. Syphilitic (H/O exposure should be taken). |
| 2. Traumatic. | 8. Marfan's syndrome (look for tall stature). |
| 3. Infective endocarditis. | 9. Ankylosing spondylitis. |
| 4. Bicuspid aortic valve. | 10. Rheumatoid arthritis. |
| 5. Atherosclerotic. | 11. Reiter's syndrome. |
| 6. Dissection of aorta. | 12. Dissecting aneurysm of aorta. |

* First 4 causes are due to 'valve involvement' and the others produce AI by 'ring dilatation' due to aortic root disease.

Causes of 'acute AI' :

Peripheral signs are absent in acutely developing AI. They develop from :

1. SBE.
2. Trauma to the chest.
3. Dissection of aorta.
4. Acute rheumatic fever.
5. Prosthetic valve dysfunction.

AI with normal or low pulse pressure :

1. Acutely developing AI (see above).

2. Associated with AS or tight MS.
3. AI with CCF.
4. AI associated with systemic hypertension.

Is there any systolic murmur present in the aortic area ?

No; occasionally a harsh systolic murmur of 'functional' AS of grade II to V may be heard in the aortic area. This murmur may be transmitted towards carotids. This is caused by increased flow across the aortic valve and does not necessarily indicate an organic AS (for the difference between organic and functional AS, read the section on Aortic stenosis and aortic incompetence').

Clinical features of 'acute AI' :

1. Acutely ill and symptomatic (breathlessness, chest pain) patient.
2. 100% patients have acute left heart failure.
3. Peripheral signs are absent (normal or low pulse pressure).
4. No cardiomegaly.
5. Diastolic thrill is present in majority of patients.
6. S_j — Soft to absent.
7. Short, soft, early diastolic murmur.

Features of rheumatic AI:

1. Age — Young age group.
2. H/O rheumatic fever present in majority of patients.
3. Involvement of other valves—May be present.
4. Diastolic thrill—Usually absent.
5. A₂ — May be diminished (soft) or absent.
6. Murmur—Best audible in left 3rd ICS (neo-aortic area), blowing in character.
7. Peripheral signs — Present.

Features of syphilitic AI:

1. Age — Older age group (> 40 years).
2. H/O exposure to syphilis—Present.
3. Usually isolated lesion. Anginal pain is more common.
4. Diastolic thrill—May be present (commoner than rheumatic).
5. Right parasternal heave—Very often present.
6. A₂ — usually loud (tambour quality).
7. Murmur — Best audible along right sternal border (right 3rd ICS); ringing, cooing or musical in nature.
8. Peripheral signs—More marked than rheumatic AI.
9. Austin Flint murmur — May be present.

* If the early diastolic murmur of AI is best audible along right sternal border, it suggests syphilis, Marfan's syndrome or ankylosing spondylitis, and is due to dilatation of ascending aorta.

** In atherosclerotic AI, the A₂ is ringing in quality.

Causes of pulsation at different areas :

(A) CAROTID PULSATION :

1. Exertion, emotion and excitement,
2. Aortic incompetence (Corrigan's sign),
3. Hyperkinetic circulatory states like severe anaemia, thyrotoxicosis, pyrexia, pregnancy etc,
4. Atherosclerosis,
5. Aortic aneurysm,
6. Kinked carotid artery (right > left).

(B) SUPRASTERNAL (JUGULAR) NOTCH PULSATION :

1. Hyperkinetic circulatory states like severe anaemia, thyrotoxicosis, pyrexia, pregnancy etc,
2. Aortic incompetence,
3. Coarctation of aorta,
4. Unfolding of the arch of aorta (old age; commonly in patient with systemic hypertension),

5. Aneurysm of the arch of aorta,
6. High arch of aorta (congenital).

(C) PULSATION OVER THE PULMONARY AREA :

- | | |
|-------------------------------------|---|
| 1. Children (physiological), | 4. PDA. |
| 2. Pulmonary hypertension (severe), | 5. VSD, |
| 3 ASD | 6. Idiopathic dilatation of pulmonary artery. |

(D) LEFT PARASTERNAL PULSATION :

1. RVH,
2. LAH (rare).
3. ASD (left parasternal lift).

(E) RIGHT PARASTERNAL PULSATION :

1. Dextrocardia,
2. Dextroversion (due to extracardiac cause, i.e., cause in lung, pleura, thoracic cage),
3. Aneurysm of the ascending aorta,
4. Right atrial hypertrophy due to any cause.

** (F) EPIGASTRIC PULSATION :*

1. Excitement specially in a thin person (physiological),
2. Right ventricular hypertrophy (RVH),
3. Transmission of aortic pulsation by any growth like gastric carcinoma (pulsation disappears after adopting knee-elbow position)—i.e., transmitted pulsation,
4. Aneurysm of the abdominal aorta (expansile pulsation),
5. Transmitted hepatic pulsation in tricuspid incompetence.

(G) PULSATION AT THE BACK :

1. Coarctation of aorta (Suzman's sign),
2. Pulmonary arteriovenous fistula.

*(H) JUXTA-APICAL : Ventricular aneurysm.**(I) APICAL : normally in health. LVH or RVH.*

* The epigastric pulsation is felt with the tip of the index finger inserted upwards and obliquely under the xiphoid process, or it is often better seen after tangential application of light.

Meaning of a 'thrill' :

This is palpable low-frequency vibrations with the sensation like 'purring of a cat', and is always associated with heart murmur. It is synonymous with palpable murmur. A thrill is always described by its site and timing.

How to determine the timing of a thrill ?

Palpate the thrill with the whole flat of the right palm (mainly by the base of the fingers.) While palpating for a thrill, always put your left thumb over the right carotid artery at the level of the upper border of thyroid cartilage to confirm the timing. If the thrill is synchronous with carotid pulsation, it is systolic in timing and if it appears before carotid pulsation, it is diastolic thrill. Many clinicians adjust the timing of a thrill with the apex beat instead of carotid pulsation.

Always remember :

- a) In mitral or apical area — Diastolic thrill is very common.
- b) In all other areas (base of the heart and tricuspid area) — Systolic thrill is more common.
- c) In pulmonary area — Thrill may be continuous or systolo-diastolic, e.g., PDA.

N.B. : It is seen that thrill is usually present in stenotic lesions and generally absent in regurgitant lesions of the heart. The presence of a thrill, most of the time, indicate that the murmur is organic.

* Sudden appearance of thrill usually indicates SBE or failure of prosthetic valve.

** Austin Flint and Carey-Coombs murmur do not have thrills.

Importance of joint involvement in CVS :

1. Acute rheumatic arthritis.
2. Rheumatoid arthritis. SLE, ankylosing spondylitis may be associated with AI or MI.
3. Arthralgia may be seen in valvular heart diseases complicated by SBE.

Why the murmur of AI is early diastolic ?

The leaking back of blood from the aorta to the left ventricle is greatest in early part of diastole for obvious reason (as pressure in the aorta is more than that of the left ventricle in early diastole). In the mid-diastole, no leak is possible as the pressure in the aorta and the left ventricle becomes equal.

Differential diagnosis (D/D) of your case ?

1. PDA, coronary arteriovenous fistula, ruptured sinus of Valsalva Considered for wide pulse pressure; they produce continuous murmur.
2. Graham Steell murmur (rare) — Considered for diastolic murmur; features of pulmonary hypertension are present without any peripheral sign of AI.

Complications of AI:

- | | |
|--------------------------------|---|
| 1. Acute LVF. | 4- Arrhythmias. |
| 2. Infective endocarditis. | 5. Heart block. |
| 3. Congestive cardiac failure. | 6. Angina pectoris (specially nocturnal). |

How do you like to investigate a case of AI ?

1. X-ray chest (PA view) :
 - a) Cardiomegaly (LVH)—also known as Cor Bovinum.
 - b) Aortic root dilatation (prominent aortic knuckle).
 - c) Aortic valve calcification (raises the possibility of concomitant AS).
2. ECG :
 - a) LVH.
 - b) Conduction defects.
3. Blood for (to detect active rheumatic carditis) :
 - a) TC and DC b) ESR c) ASO titre, and
 - d) VDRL and Kahn test (for syphilis); ANF, rheumatoid factor, CRP to exclude connective tissue diseases.
4. Echocardiography (M-mode and 2-D)—to diagnose, assess severity and possible cause of lesion.
5. Cardiac catheterisation.
6. Doppler study—most sensitive for detecting mild AI.

How will you treat this case ?

1. Medical management - as in MS; underlying cause, e.g., infective endocarditis or syphilitic aortitis needs to be treated. Chronic AI with significant volume overload needs vasodilator therapy with hydralazine, nifedipine or ACE-inhibitors.
2. Surgical — aortic valve replacement (prosthesis); ideal time of operation is selected after haemodynamic or echocardiographic evidence of left ventricular dysfunction but before the development of significant symptoms.

N.B. : Diastolic thrill in the aortic area is rare in AI. If really a diastolic thrill is present, it is better to think of leucic origin or ruptured sinus of Valsalva or AI developed from dissecting aneurysm.

Common causes of palpitation :

Palpitation means 'abnormal unpleasant awareness of the heart beat.

(A) Cardiac —

- | | |
|-----------------------------------|--------------------------|
| 1 AI, MI, TI, MS. | 4- Atrial fibrillation. |
| 2 ASD VSD, PDA. | 5. Ventricular ectopics. |
| 3. Paroxysmal atrial tachycardia. | 6. Brady arrhythmias. |

(B) Non-cardiac —

1. Hyperkinetic circulatory states like severe anaemia, thyrotoxicosis, arteriovenous shunt, anxiety, pyrexia.
2. Drug-induced, e.g., sympathomimetics (e.g., salbutamol), vasodilators (e.g., nifedipine), atropine.
3. Non-organic diseases, e.g., excessive smoking, anxiety states, cardiac neurosis, hysteria.
4. Miscellaneous—Hypoglycaemia, pheochromocytoma, menopause.

Case 5

MITRAL STENOSIS AND MITRAL INCOMPETENCE

What is your diagnosis ?

This is a case of organic mitral incompetence and mitral stenosis of rheumatic origin with pulmonary hypertension but without any evidence of congestive cardiac failure or arrhythmia at present (start with the dominant lesion).

N.B. . Never use the term double mitral lesion or MS with MI (abbreviations) in the examination. A combined lesion is difficult to be diagnosed correctly by an undergraduate student. But if one is meticulous and to the point in the clinical examination, it is not very difficult to diagnose the lesion. In a patient with MS and MI, both may be organic or the MS may be functional, if present with gross MI. When both are organic lesions, it is the duty of a student to determine the predominant (haemodynamically significant) one.

Palpatory findings in the apical area in this patient :

1. Site of the apex beat is in left 6th ICS, 1/2" outside the left MCL (LVH due to MI).
2. Character—Hyperdynamic (due to MI).
3. Palpable S_t —Absent.
4. Thrill Systolic thrill, best felt in left lateral position with full expiration.
Here, all the features point towards predominant MI.

N.B. : Remember, diastolic thrill in MS is more common than systolic thrill in MI (in the mitral area).

If MS is the predominant lesion, the findings in the apical area would be :

1. Site—Left 5th ICS, 1/2" outside the left MCL (RVH).
2. Character—Tapping.
3. Palpable S_t —Present.
4. Thrill Diastolic thrill, best felt in left lateral position with full expiration.

Is pulmonary hypertension present here ?

Yes. In a combined lesion of MS and MI, pulmonary hypertension is usually present though it depends mainly on the duration of the disease.

How to determine the dominant lesion ?

FEATURES	MS>MI	MI>MS
(A) Symptoms :		
Haemoptysis, H/O PND, systemic embolisation, symptoms of CCF, pulmonary congestion, chest pain	+++	+
(B) Signs :		
1. Pulse—	Low volume, features of atrial fibrillation are more common	Within normal limit (not low volume); may be 'collapsing'
2. BP—	Low systolic BP	Within normal limit
3. Apex—	Already described	Already described
4. Left parasternal heave—	+++	+
5. S_1 —	Short, sharp and accentuated (snapping)	Soft or muffled
6. Opening snap—	Usual and the lesion is organic	OS is absent in a dominant MI
7. S_3 —	Absent	+; Presence of S_3 signifies dominant and gross MI

FEATURES	MS>MI	MI>MS
8. Diastolic murmur—	Classical murmur of MS	Short mid-diastolic flow murmur without presystolic accentuation (due to functional MS)
9. Systolic murmur—	Pansystolic blowing murmur of MI without radiation	Classical pansystolic murmur of MI with radiation
(C) Investigations :		
1. Chest X-ray (PA view)—	LAH + RVH	LVH + LAH
2. ECG—	See the section on 'MS' (LVH is not present)	See the section on 'MI' (LVH is usually present)
3. Echocardiography—	Final diagnosis with certainty (thick, immobile cusps with diminished rate of diastolic filling)	Final diagnosis with certainty (LA enlargement with hyperdynamic LV)

* MS>MI → commonly dyspnoea on exertion → orthopnoea → PND → haemoptysis; palpitation in the presence of AF.

** MI>MS → commonly palpitation → dyspnoea → orthopnoea → PND.

What are the evidences of MI present here ?

Read the differentiation carefully and give your answer accordingly.

Why this is MI murmur, not TI ?

Describe the classical MI murmur. It is best heard over the mitral area with typical radiation, and increases with expiration. The TI murmur is best heard in lower left parasternal area, localised and increases with inspiration (**Carvallo's sign**).

Conditions associated with S_3 :

S_3 , signifies diastolic overload of the ventricles. It occurs during the first rapid filling phase of cardiac cycle (in ventricular diastole). It is a ventricular distension sound and the clinical associations are :

1. Healthy young adults (< 40 years) and children, pregnancy, athletes.
2. Heart failure (LVF or RVF).
3. Chronic MI, TI, AI.
4. Hyperkinetic circulation like severe anaemia, thyrotoxicosis.
5. ASD. VSD or PDA; dilated cardiomyopathy.

Conditions associated with S_4 :

It signifies systolic overload of the ventricles. It occurs during the last rapid filling phase of cardiac cycle (in ventricular diastole) which also coincides with the atrial systole. Presence of S_4 is always abnormal. It is an atrial contraction sound and the clinical associations are :

1. Ischaemic heart disease, specially acute myocardial infarction.
2. Aortic stenosis.
3. Systemic hypertension.
4. Rarely, in normal individuals (above 60 years).
5. Hypertrophic cardiomyopathy.

How to auscultate for S_3 or S_4 ?

S_3 or S_4 is best heard at the **apex** with the **bell** of stethoscope placed lightly. Sometimes, they are best heard with the patient turned to **left lateral** position; often they are better felt than heard. They are low-pitched sounds.

Left-sided S_3 (LVF) is best audible at the apex during expiration while the right-sided S_3 (RVF) is best heard at the lower left sternal border during inspiration.

Can you accentuate gallop sounds ?

Elevate both lower limbs at 60° → maintain the position for 30 seconds → cardiac output ↑ →

accentuation of gallop sounds. In this way a latent S_3 can be made audible. Isometric handgrip also accentuate gallop.

What is dynamic auscultation ?

Circulatory dynamics can be altered by application of certain body manoeuvres (respiration, isometric handgrip, Valsalva manoeuvre, leg raising) and administering certain vasoactive substances (amyl nitrite, phenylephrine), and auscultation of these events in the CVS is known as dynamic auscultation.

Table 2 : Dynamic manoeuvres in differentiation of murmurs

Manoeuvres	Increased	Decreased
1. Isometric handgrip (T afterload)	AI, MI	HOCM, MVPS
2. Valsalva manoeuvre (i venous return)	HOCM, MVPS	All organic murmurs except HOCM and MVPS
3. Squatting (T venous return and T peripheral resistance)	AS, AI, MI	HOCM, MVPS
4. Standing (l venous return)	HOCM, MVPS	AS, MI
5. Both leg raising (T venous return)	All right-sided murmur	HOCM, MVPS
6. Inspiration (T venous return)	All right-sided murmur	Pulmonary ejection click
7. Expiration (T cardiac output of left heart)	All left-sided murmur	—
8. Amyl nitrite (J. arterial pressure and T cardiac output)	HOCM, AS, MS	AI, MI

HOCM (hypertrophic obstructive cardiomyopathy), MVPS (mitral valve prolapse syndrome)

How and where the 'bell' is used ?

During the use of the bell, it should be placed very lightly over the skin. More pressure will tighten the skin and smother the low-frequency sounds. It is made for listening low-pitched sounds like,

1. Murmur of MS and TS.
2. S_3 or S_4 .
3. Foetal heart sounds.
4. Venous hum.

Cardinal points to diagnose dominant lesion (MS + MI) :

1. History.
2. Pulse.
3. Apex (LVH or RVH, hyperdynamic or tapping).
4. S_1 (loud or muffled).
5. S_3 (S_3 signifies significant MI).
6. Opening snap (always signifies dominant MS).
7. Mid-diastolic rumbling murmur with presystolic accentuation signifies predominant MS; only mid-diastolic murmur points towards a dominant MI.

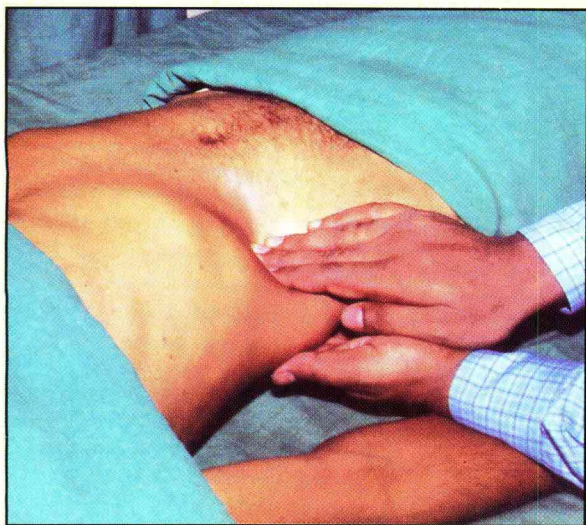
Mitral diastolic murmur—organic or functional ?

Functional MS (from gross MI) will have the following features :

1. Pulse—Normal volume.
2. Diastolic thrill in mitral area—Absent; rather a systolic thrill may be present.
3. S_1 —Not very loud.
4. Presystolic accentuation (of the murmur) is absent.
5. Opening snap—Absent; rather S_3 is heard due to significant MI.
6. Features of LVH.

D/D of opening snap :

1. Splitting of S_2 , 2) S_3 , and 3) Pericardial knock.



Bimanual **palpation** of right kidney



Elicitation of **fluid thrill**



Examination for '**fluctuation**' at nail-bed



Measurement of **jugular venous pressure (JVP)**



Examining '**pulse deficit**' simultaneously by two examiners



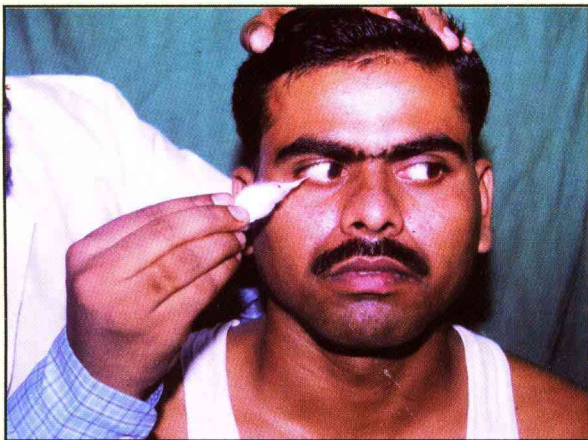
Eliciting **flapping tremor**



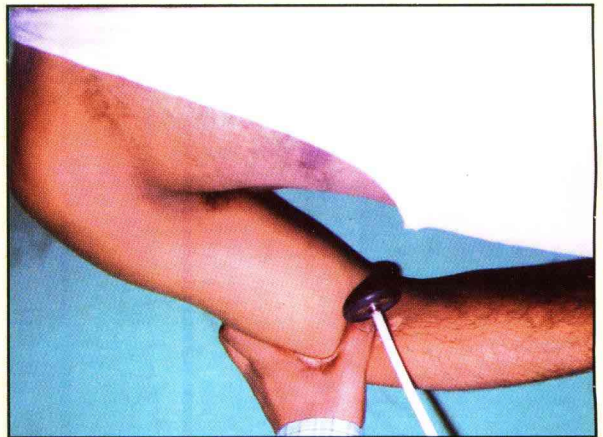
Testing for **neck rigidity**



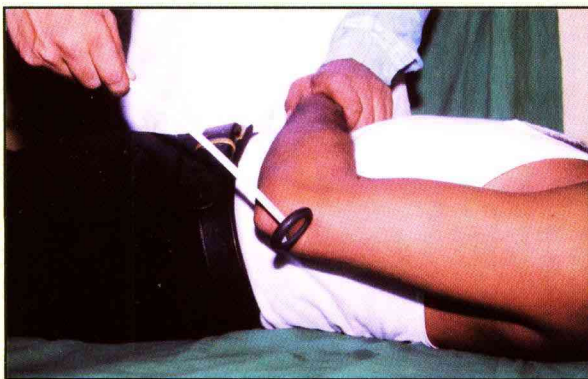
Eliciting **Kernig's sign**



Demonstrating **corneal reflex**



Testing **biceps jerk**



Testing **triceps jerk**



Testing **supinator jerk**

Table 3 : Differentiation between opening snap and S₃

Features	Opening snap	S ₃ (LV)
1. Where found —	MS	Heart failure or chronic MI
2. Best audible—	At lower left parasternal region; with the diaphragm of stethoscope in standing position	At apex; with the bell of stethoscope in left lateral position
3. Pitch—	High-pitched	Low-pitched
4. Palpability—	Not palpable	May be palpable
5. Timing—	0.04 to 0.12 seconds after A ₂ (early part of diastole)	0.14 to 0.16 seconds after A _y (mid-.diastole)
6. After treating heart failure—	OS becomes louder	S ₃ vanishes

What is the aetiology of your case ?

Rheumatic; an organic MS with MI is virtually always of RHEUMATIC in origin.

How will you manage this case ?

1. Prophylaxis against rheumatic recurrence.
2. Mitral valve replacement (prosthesis).
3. Treatment of complications :
 - a) Heart failure, b) SBE, c) Atrial fibrillation, d) Embolic manifestations.

Case 6**AORTIC STENOSIS AND AORTIC INCOMPETENCE****What is your diagnosis ?**

This is a case of organic aortic incompetence and aortic stenosis of rheumatic origin without any evidence of congestive cardiac failure and the patient is in sinus rhythm at present (start with the dominant lesion).

N.B. : Do not use the term 'double aortic' lesion or AS with AI (abbreviations) in the examination. Often a functional systolic murmur is heard in a patient with severe AI. Thus, one has to determine whether the AS is organic or functional. Lastly, in a combined AS with AI, dominant lesion (haemodynamically significant) should be diagnosed. It is worthwhile to remember that combined AS with AI is usually of rheumatic in origin.

How to determine the dominant lesion ?

FEATURES	AS>AI	AI>AS
(A) Symptoms :		
1. Effort angina, dyspnoea on effort, effort syncope, black-out etc	+++	+ (chest pain, dyspnoea may occur)
2. Palpitation, throbbing	+ (palpitation)	+++
(B) Signs :		
1. Pulse—	Low volume; pulsus bisferiens (tidal>percussion wave)	High volume collapsing pulse; pulsus bisferiens (percussion> tidal wave)
2. BP—	Low systolic BP; pulse pressure is low	Wide pulse pressure due to high systolic and low diastolic BP

FEATURES	AS>AI	AI>AS
3. Peripheral signs of AI—	±	+++
4. Apex—	Heaving, apex may be in normal position	Hyperdynamic, down and outwards (LVH)
5. Thrill—	Always systolic; palpated on aortic area and on carotids	a) Rarely present; systolic thrill due to functional AS b) Very rarely diastolic thrill may be palpable along the left sternal border
6. S₃-	Absent	May be present
7. S₄-	May be present	Absent
8. Ejection click—	Present in mild to moderate AS	Rare; sometimes present in severe AI
9. Diastolic murmur—	Very short duration AI murmur	Classical murmur of AI
10. Systolic murmur—	Classical murmur of AS	Functional midsystolic ejection murmur may be audible

(C) Investigations :

- | | | |
|---------------------------|--------------------------------|--------------------------------|
| 1. Chest X-ray (PA view)— | Read the section on AS' | Read the section on AI' |
| 2. ECG— | Read the section on AS' | Read the section on AI' |
| 3. Echocardiography— | Final diagnosis with certainty | Final diagnosis with certainty |

Cardinal points to diagnose dominant lesion (AS + AI) :

- | | |
|---|--|
| 1. History — angina and syncope are seen more in AS. Palpitation and dyspnoea are common in AI. | 4. Apex. |
| 2. Pulse. | 5. S ₃ , or S ₄ . |
| 3. BP. | 6. Ejection click. |
| | 7. Peripheral signs of AI (best indicator of severity of AI lesion). |

Aortic stenosis murmur—organic or Junctional ?

- History—Patients with organic AS will give H/O angina and/or syncope.
- Pulse— Low volume, often anacrotic in nature (in organic AS).
- BP—Systolic decapitation of BP in organic lesion.
- Apex—Heaving in organic lesion.
- Systolic thrill in aortic area—Present in organic lesion (rarely, functional AS may give rise to systolic thrill).
- Ejection click—Always in favour of valvular (organic) AS.

* In a functional AS, peripheral signs of AI will be very prominent like presence of water-hammer pulse, dancing carotids etc. Thus in a functional AS, pulse is of high volume.

Possible BP in patients where AS>AI and AI>AS :

If AS>AI — the approximate BP will be 90/60 mm of Hg and if AI>AS — the approximate BP will be 130/20 mm of Hg. So, measurement of BP should be done carefully in all CVS patients.

What is aortic sclerosis murmur ?

In aged persons suffering from atherosclerosis with or without hypertension, there is fibrosis, thickening and calcification of bases of the aortic valve cusps. These may give rise to harsh, ejection systolic murmur upto grade IV in some cases. Differentiate aortic sclerosis from aortic stenosis by,

- Normal pulse volume,
- Absence of thrill.
- Normal A₂, and
- Often by the features of atherosclerosis like thick peripheral arteries, kinked carotids, locomotor brachialis, suprasternal pulsation, xanthelasma (deposition of lipids) around the eyes. Occasionally, the calcification becomes excessive and thus, produces severe aortic valve obstruction. This is known as 'calcific aortic stenosis' (see aetiology of AS).

What is the possible aetiology of your case?**Rheumatic.**

Sometimes, the bicuspid aortic valve (congenital) produces features of combined AS and AI though the AI lesion is insignificant (always AS>AI). Bicuspid aortic valve is the most common congenital malformation of the heart though it may go undetected in early life.

Signs of aortic arch aneurysm :

1. Swelling in the chest wall.
2. Pulsation in the upper part of anterior chest wall.
3. Suprasternal pulsation.
4. Difference between right and left radial pulses (radio-radial delay).
5. Left 2nd ICS is dull on percussion.
6. Ejection click.
7. Systolic murmur in the aortic area.
8. 'Tracheal tug'— It is the downward pull of trachea synchronous with each cardiac cycle. Stand in front of the erect (upright) patient. Raise the chin and apply upward pressure on the trachea with two fingers placed on the lower border of cricoid cartilage. A downward traction is felt on the trachea. This sign is also seen in COPD patients when the downward tracheal pull is felt on each inspiration. The other name of this manoeuvre is **Oliver's sign**.

Differential diagnosis (D/D) of your case :

Actually, a patient with AS and AI having systolic and diastolic murmur in aortic area mimic a 'continuous murmur'. The common causes of **continuous murmur** (begins in systole and extends through second heart sound into part or whole of diastole) are :

1. Patent ductus arteriosus (PDA, Train in tunnel' murmur, machinery murmur, Gibson's murmur)—Commonly found in female patients with water-hammer pulse and continuous murmur with late systolic accentuation, best audible in left infraclavicular area and left 2nd ICS. Thrill may be systolic or systolo-diastolic.
2. Rupture of sinus of Valsalva—Dramatic onset of chest pain with continuous murmur along the left sternal border.
3. Coronary arteriovenous fistula—Along left sternal edge, superficial, harsh.
4. Pulmonary arteriovenous fistula— Along left 2nd ICS.
5. Aortopulmonary window— Along left 3rd ICS.
6. Rarely, in coarctation of aorta (better auscultated at the back).
7. Mammary souffle (in pregnancy) —Best heard over the mammary area, and 2nd to 4th ICS.
8. Venous hum at root of neck (Devil's murmur) — Actually it is a D/D of continuous murmur.
9. VSD with AI, AS with AI, and MS with MI mimic continuous murmur.
10. After Blalock-Taussig operation done in Fallot's tetralogy— In upper sternal area.

Continuous murmur with high pulse pressure :

1. PDA, 2. Rupture of sinus of Valsalva, and 3. Aortopulmonary window.

Clues to diagnose multivalvular lesions :

1. Mild or moderate AI is frequently missed. Carefully listen to the aortic and then the neoaortic area (Erb's point) in sitting posture of the patient, in full expiration and with the diaphragm of stethoscope, in the search of an early diastolic murmur. For confirmation, measure the BP (in all cases) to record the pulse pressure.
2. Short mid-diastolic murmur of MS is often missed. Thus, it is a dictum to listen the mitral area with the bell of stethoscope, in left lateral position and with full expiration. For corroboration, carefully auscultate for loudness of S .
3. Murmur of insignificant MS may be pronounced after little exercise, if the condition of the patient permits (e.g., in the absence of heart failure).
4. Carefully palpate the brachial and carotid arteries. Actually, the commonest cause of pulsus bisferiens is combined lesion of AS and AI. If you get pulsus bisferiens in your patient, you are sure about the presence of two valvular diseases (rarely, it is seen in isolated AI).

Causes of LVF :

1. Systemic hypertension.
2. IHD, i.e., acute myocardial infarction.
3. AI.
4. AS.
5. MI.
6. Cardiomyopathy.
7. Coarctation of aorta.
8. Myocarditis.
9. Severe anaemia.
10. VSD, PDA.

Causes of LAF :

1. Mitral stenosis.
2. Left atrial myxoma.
3. Ball valve thrombus in the left atrium.
4. Mitral atresia (rare).

* **Bernheim effect** (it is the RV pressure change resulting from LVH without developing pulmonary hypertension. Actually, interventricular septum hypertrophies from LVH and produces obstruction to right ventricular outflow and thus, manifests as prominent a-wave in the neck vein without developing RVH or RVF). Lungs are clear on auscultation.

** Commonest cause of RVF is LVF.

Causes of RVF (CCF) :

1. Secondary to left-sided heart failure (LVF/LAF).
2. COPD.
3. Pulmonary stenosis.
4. ASD, VSD or PDA.
5. Pulmonary hypertension due to any cause.
6. Pulmonary thromboembolism.
7. Cardiomyopathy.
8. Myocarditis.
9. Right ventricular infarction.

Causes of RAF:

1. Tricuspid stenosis.
2. Secondary to RVF.
3. Right atrial myxoma (very rare).

Causes of biventricular failure :

1. Hyperkinetic circulation, e.g., severe anaemia, thyrotoxicosis, arteriovenous shunts.
2. Myocarditis—Viral, rheumatic, diphtheritic etc.
3. Ischaemic heart disease.
4. Dilated cardiomyopathy.
5. Patients with systemic hypertension and pulmonary hypertension (commonly from COPD).
6. RVF following chronic left-sided heart failure.

Causes of sudden cardiac failure :

1. Acute myocardial infarction.
2. Malignant or accelerated hypertension.
3. Dissection of aorta.
4. Massive pulmonary thromboembolism.
5. Arrhythmias.
6. Circulatory overload (fluid infusion or blood transfusion, severe anaemia).
7. Myocarditis.
8. Subacute bacterial endocarditis.

Causes of high output failure :

1. Hyperthyroidism.
 2. Severe anaemia.
 3. Arteriovenous fistula.
 4. Paget's disease.
 5. Pregnancy
 6. Chronic cor pulmonale.
 7. Beriberi.
- * In high output failure, cardiac output is usually higher than normal.

Accelerated and malignant hypertension :

(A) Accelerated hypertension—Significant recent increase in BP level associated with evidence of vascular damage on fundoscopic examination but **without papilloedema**.

(B) Malignant hypertension—Severe accelerated hypertension with diastolic BP > 130 mm of Hg. with haemorrhage, exudate and **papilloedema** found on fundoscopic examination, and often associated with one or more of the following :

- a) Rapidly deteriorating renal function (oliguria).
- b) Cardiac decompensation (acute LVF).
- c) Central nervous system manifestations, e.g., hypertensive encephalopathy (severe headache, vomiting, convulsions, coma).

It is papilloedema, not the diastolic BP—which is important here.

Management of multivalvular heart diseases :

1. AS with AI is managed by aortic valve replacement (prothesis).
2. MS with AI—First the AI is dealt with and next the MS. If MS is operated before AI, there will be left ventricular overload (blood coming from LA and aorta both) and ultimately LVF occurs. Actually, both the operations are done simultaneously.
3. MS with AS — MS protects the heart from failure. First deal with AS and then MS.

Case 7

INFECTIVE ENDOCARDITIS

What is your diagnosis ?

This is a patient of rheumatic mitral incompetence complicated by subacute bacterial endocarditis (SBE) and without any heart failure or dysrhythmia at present.

Reasons behind your diagnosis :

(A) Symptoms (from the history) —

Fever with chills present persistently with diaphoresis, arthralgia, lassitude, anorexia, pain in the loin etc; over and above dyspnoea, fatigue, chest discomfort and palpitation due to MI.

(B) Signs —

1. Anaemia — Severe with a peculiar pallor (cafe-au-lait pallor).
2. Temperature (oral) — Elevated (101°F); toxic look.
3. Clubbing — Present (may be painful).
4. Pulse — Tachycardia, regular in rhythm (describe all points).
5. Petechiae, Osier's node and splinter haemorrhage — Present.
6. Pansystolic murmur (due to pre-existent MI) with a musical quality — 'Seagull murmur'.
There may be change in the pre-existent murmur or appearance of a new diastolic murmur.
7. Mild, tender splenomegaly (30-40%) with splenic rub.
8. Renal angle is tender.
9. Arthralgia or arthritis.
- [10. Ophthalmoscopy — Roth spots in <5% cases (see page 53)].

Hands in SBE :

1. Clubbing (10%).
2. Pallor (due to anaemia).
3. Osier's node (5%)—Painful and tender papule about the size of pinhead to pea, and is seen in the *pulp offingers*, toes and palms (due to septic embolism or arteritis). The nodes usually last for 1-5 days.
4. Splinter haemorrhage (10%; also known as Horder's line)—Linear longitudinal haemorrhage under the nail (other causes of splinter haemorrhage are trauma, trichinosis, leukaemia, scurvy, psoriasis, rheumatoid arthritis, systemic vasculitis).
5. Janeway's spot — Large, non-tender maculopapular lesion *in palms* (and soles).
6. Temperature — Elevated.
7. Petechiae (40-50%) may be found in dorsum of hands.
8. Gangrene of the fingers due to embolism rarely.
9. Nail-fold thrombi (due to embolisation).
10. Painful finger tips; loss of pulses (embolic manifestation).

* 3, 4, 5, 7, Roth spots, nephritis and positive rheumatoid factor are immunological phenomenon.

** Features of septic embolisation : 3, 4, 5, 7-10, painful splenomegaly, CNS and renal manifestations.

*** Though initially thought of an exclusive manifestation of SBE, Roth spots are also non-specific sign in blood dyscrasias (e.g., acute leukaemias) and anaemia (e.g., aplastic anaemia).

D/D you like to consider in your patient:

The features like fever, cardiac murmur and embolic phenomenon may be present in,

- | | |
|---------------------------|----------------------------------|
| 1. Acute rheumatic fever, | 3. Marantic endocarditis, |
| 2. Atrial myxoma (left), | 4. Systemic lupus erythematosus. |

The clinical findings in different systems in SBE :

1. General—Pyrexia, malaise, anorexia, diaphoresis, weight loss.
2. CVS—Breathlessness, pain chest, palpitations, tachycardia, changing of pre-existent murmur or appearance of new murmur, CCF and conduction defects.
3. Kidneys—Haematuria, glomerulonephritis, loin pain due to embolic renal infarction, tender renal angle.
4. Lungs—Haemoptysis, chest pain, pleural rub (embolic infarction of lung).

5. Nervous system—Headache, meningitis, embolic stroke, convulsions, intracranial haemorrhage due to ruptured mycotic aneurysm, encephalopathy.
6. Blood vessels—Cold periphery due to loss of pulses as a result of embolisation.
7. Skin and mucous membrane—Petechiae, purpura, subungual haemorrhage.
8. Hands—Described above.
9. Eyes—Red eyes, sudden blindness, subconjunctival haemorrhage, Roth spots.
 10. Haematopoietic—Fatiguability, anaemia, leucocytosis, thrombocytopenia, T ESR and T CRP.
 11. Spleen—Splenomegaly, pain in left hypochondrium, splenic rub (due to splenic infarction).
11. Bowel—Pain abdomen, mesenteric artery embolism, bowel infarction.

* Read modified Duke criteria for the clinical diagnosis of infective endocarditis from any standard text book.

How to establish the diagnosis of SBE ?

1. Blood—
 - a) Anaemia (normocytic-normochromic).
 - b) Leucocytosis (neutrophilia).
 - c) Thrombocytopenia.
 - d) Raised ESR and high C-reactive protein.
 - e) Blood biochemistry—urea, creatinine, bilirubin and alkaline phosphatase levels may be elevated.
 - f) Immunoglobulins and complements—tserum immunoglobulins, Tcirculating immune complexes and ^-complement levels are characteristic.
 - g) Repeated blood culture (typical organism for infective endocarditis are diagnosed from two seperate blood cultures drawn 12 hours apart which detect bacteraemia in 90% culture-positive cases; or detected from all of 3, or majority of 4 or more seperate blood cultures done, with the first and last blood cultures obtained at least 1 hour apart. Blood is drawn under aseptic technique from different venepuncture sites; both aerobic and anaerobic, and rarely fungal cultures are done). At least 10-20 ml blood is taken at a time which is not related to height of temperature. An indwelling I.V line should not be used for collection of blood. *Streptococcus viridans* is the commonest pathogen in SBE. Positive blood culture gives the definite diagnosis of SBE. Presumptive antibiotic therapy should be started immediately after obtaining the blood culture samples.
2. Urine—
 - a) Microscopic haematuria (frank haematuria is rare).
 - b) Slight proteinuria.
3. Chest X-ray—cardiac failure, cardiomegaly.
4. Electrocardiography (ECG) —

Sinus tachycardia, prolonged PR interval, AV block (if septum is involved).
5. Echocardiography (2-D) — to assess valve damage, identification of vegetations (small/sessile/polypoidal), progress of vegetations, and abscess detection :
 - a) Demonstration of parent disease (e.g.. MI here) with chamber dilatation.
 - b) Vegetations (> 2 mm by transthoracic echo and < 2 mm by transoesophageal echo) are detected.
6. Bone marrow culture and serological study for Candida, Histoplasma and Brucella may be helpful in culture-negative cases.

* Positive past H/O recent dental extraction, genitourinary instrumentation or IV drug abuse is important in diagnosis of SBE.

Haematuria in SBE :

1. Autoimmune nephritis, and/or
2. Infarction of the kidney.

Features in the nervous system (in SBE) :

They are due to embolism or mycotic aneurysm. The features are,

1. Toxic psychosis, meningitis, metastatic brain abscess.
2. Convulsions, hemiplegia, intracranial haemorrhage due to haemorrhagic infarcts, subarachnoid haemorrhage (mycotic aneurysm), coma.

3. Roth spots—Yellowish, elliptical, *flame-shaped*, *haemorrhage with a pale centre* present in the retina and is found in ophthalmoscopy. This is due to deposition of circulating immune complex (other causes of Roth spots are aplastic anaemia, leukaemia, scurvy, dysproteinaemia and HIV retinopathy).

Right-sided endocarditis :

1. *Commonest organism is Staphylococcus aureus.*
2. Affects mainly the tricuspid valve and rarely the pulmonary valve.
3. Common in drug addicts or patients receiving central venous catheters.
4. Prognosis is relatively better.
5. Systemic embolisation is rare though pulmonary infection and infarction are not uncommon.

Clinical spectrum of infective endocarditis :

It is of two types :

1. Acute bacterial endocarditis—see below.
2. Subacute bacterial endocarditis (SBE)—caused by less virulent organisms with an indolent course. Cardiac damage occurs slowly and rarely metastasize unless complicated by a major embolic event. Organisms are *Streptococcus viridans* (commonest), staphylococci, HACEK organisms (*Haemophilus*, *Actinobacillus*, *Cardiobacterium*, *Eikenella* and *Kingella*), fungi (*Candida*) or rickettsia. SBE affects a patient who is suffering from either rheumatic heart disease or congenital heart disease or has undergone some sort of cardiac surgery (e.g., valvotomy, prosthesis).

Acute bacterial endocarditis :

1. Often involves the normal heart with a rapidly progressive course (SBE invariably involves damaged heart), and is caused by highly virulent and invasive organisms.
2. Right-sided involvement (tricuspid valve) is common; pneumonia may be present.
3. Clubbing is not a feature.
4. Common in I.V drug abusers.
5. Source of CVS involvement is often evident i.e., there may be preceding staphylococcal abscess or pneumococcal meningitis. *Staphylococcus aureus* is the commonest pathogen.
6. Cardiac and renal failure develop rapidly.
Rapid onset and fulminant course; damages cardiac structures very rapidly and haematogenously metastasize in extracardiac sites.

Causes of culture-negative SBE :

This accounts for 5-10% of cases.

1. Partially treated patients.
2. Fungal, yeast infection or Q fever.
3. Intermittent bacteraemia.
4. L-form (cell-wall deficient form) of organism or fastidious organism (e.g., *H. parainfluenzae*).
5. Marantic endocarditis (non-bacterial thrombotic endocarditis, usually in elderly dying patients with advanced carcinomatosis).
6. Libman-Sacks endocarditis (non-bacterial verrucous endocarditis in SLE).
7. Right-sided endocarditis.

* 5 and 6 are examples of non-infective endocarditis.

Poor prognosis in SBE :

- | | |
|--|--|
| 1. Prosthetic valve, fungal or gram-negative endocarditis. | 3. Presence of congestive cardiac failure, |
| 2. Culture-negative endocarditis. | 5. Polymicrobial bacteraemia. |
| 4. Involvement of multiple valves. | |

Cardinal features of SBE for 'bedside' diagnosis :

- | | |
|---|--------------------------------|
| 1. Fever (persistent or swinging), toxic look | 4. Mild splenomegaly, |
| 2. Pallor, | 5. Microscopic haematuria, and |
| 3. Clubbing, | 6. Embolic manifestations. |

* As soon as SBE is diagnosed, it should be managed very promptly. The life of the patient is in danger if there is delay in the diagnosis and initiation of treatment. **So, it is a rule to examine all CVS patients for anaemia, pyrexia, clubbing and splenomegaly.**

SBE with absence of fever :

1. CCF.
2. Uraemia.
3. Fungal endocarditis.
4. Severe sepsis.
5. Prior antibiotic therapy.
6. Elderly patients.

Cardiac diseases predisposing SBE :

1. Mitral and aortic valve diseases (mainly regurgitant diseases as regurgitant jet damages the endocardium and predisposes a bed for seeding of the microorganism) i.e., MI, mitral valve prolapse, AI commonly; MS, AS rarely.
2. VSD, PDA, bicuspid aortic valve, Fallot's tetralogy, rarely ASD.
3. Prosthetic heart valves, indwelling catheters.
4. Prior cardiac surgery (valvotomy, valve replacement).

N.B. : Read the management of CCF, LVF, atrial fibrillation, rheumatic fever and SBE in details from standard text books.

O

Section 2**RESPIRATORY SYSTEM****The cardinal symptoms of respiratory system are :**

1. Cough
2. Expectoration
3. Haemoptysis (expectoration of blood or bloody sputum)
4. Chest pain (pleural pain or pain of tracheitis)
5. Breathlessness
6. Wheeze or stridor

Other symptoms are,

7. Hoarseness of voice
8. Heaviness in the chest
9. Fever
10. Swelling of feet (in chronic cor pulmonale).
11. Epistaxis (nosebleeds), symptoms related to ear/nose/throat (e.g., sinusitis, rhinitis, running nose, sneezing)
12. Syncope (e.g., cough syncope), bone pain (hypertrophic osteoarthropathy or rib pain in bronchogenic carcinoma), altered mentation (respiratory failure)
13. General symptoms — Appetite, loss of weight (pulmonary tuberculosis or bronchogenic carcinoma), fatigue, sleep, bladder and bowel

* **Upper respiratory tract** extends from external nares to the junction of larynx with trachea (i.e., upto lower border of cricoid cartilage), and includes nasal cavity, nasopharynx, sinuses, oropharynx and larynx. **Lower respiratory tract** includes trachea, lobar bronchus, segmental bronchi, generations of tracheo-bronchial tree and alveolar air sacs.

Scheme of Examination**(A) Upper respiratory tract (URT) :**

1. Nose (congestion, discharge, hypertrophied turbinate, bleeding spots and polyp) and nasal septum (deviation); alae nasi (nasal flare).
2. Mouth breathing (e.g., adenoids) or purse-lip respiration (e.g., COPD).

3. Air sinuses (examine for tenderness over maxilla; ask the patient to sit and lean forward whether any heaviness on forehead is complained or not).
4. Pharynx (throat, gum, teeth, palate, posterior pharyngeal wall, post-nasal drip), nasopharynx, tonsils with oral hygiene; halitosis present or not
5. Larynx—Could not be examined (examined by direct and indirect laryngoscopy).

* The motto of examination of URT is to determine whether it is healthy/unhealthy, or favouring aspiration or not.

(B) Examination of the chest (lower respiratory tract) :

I. INSPECTION :

1. Shape of the chest.
2. Movement of the chest.
 3. Apical impulse (stand on the right side of the patient and look tangentially over precordium).
4. Respiration — Rate, rhythm, type, depth and breathing pattern.
5. Venous prominence.
6. Fullness or depression —
 - (i) Unilateral or bilateral.
 - (ii) Localised or generalised.
7. Supraclavicular and infraclavicular fossae.
8. Whether both the nipples are at the same level or not.
9. Whether accessory muscles of respiration are working or not.
10. Wheeze or stridor.
11. Intercostal suction.
12. Skin (front and back) —

Gynaecomastia (from bronchogenic carcinoma), pigmentation, sinus (e.g., from empyema, cold abscess or systemic fungal infection), ulcer, herpes zoster, puncture mark, any swelling (e.g., cold abscess or metastatic nodule), parietal oedema (e.g., amoebic liver abscess, empyema thoracis), shiny skin over empyema thoracis, blue markings for radiotherapy over sternum etc.
13. BACK —

Scoliosis, kyphosis, drooping of the shoulder, winging of the scapula, gibbus, position of inferior angle of both scapula, symmetry of interscapular areas (spino-scapular distance), ankylosing spondylitis and venous prominence. Also observe the skin condition (e.g., sinus) over back.

* Any deformity after thoracic operation e.g., thoracoplasty, should be carefully looked for.

II. PALPATION :

1. Surface temperature.
2. Tenderness (rib tenderness in trauma, fracture or secondary metastasis; intercostal tenderness or punch tenderness in liver abscess or empyema thoracis).
3. Corroboration of the findings of inspection—Examine the spinal deformity, direction of venous flow, details of any swelling present, parietal oedema etc.
4. Position of the trachea and the apex beat (i.e., position of the mediastinum).
5. Movement of the chest (always measure the expansion of the chest by a tape in full inspiration and in full expiration).
6. Vocal fremitus.
7. Any other palpatory findings i.e., 'palpable accompaniments' like friction fremitus, rhonchial fremitus, palpable coarse crepitations, subcutaneous emphysema, palpable pericardial rub.

III. PERCUSSION ;

	Right	Left
a) Conventional percussion		
b) Shifting dullness	—	
c) Coin percussion		
d) Hepatic and cardiac dullness — Lost or not (both are lost in emphysema); delineate the upper border of hepatic dullness along the right MCL.		

IV. AUSCULTATION :

	Right	Left
a) Breath sounds	—	—
b) Vocal resonance	—	—
(diminished or increased; whispering pectoriloquy, aegophony)		
c) Adventitious sounds	—	—
(rhonchi, crepitations and pleural rub)		
d) Miscellaneous	—	—
(succussion splash, post-tussive suction etc.)		

* It is better to auscultate all over the chest without leaving any space unexamined.

** Never forget to examine the upper respiratory tract and the back in respiratory system.

*** **No examination of the lungs, however carefully performed, should be considered entirely satisfactory or final, if made in the recumbent position.**

Case 8

PLEURAL EFFUSION

What is your diagnosis ?

This is a case of left-sided massive pleural effusion, probably of tuberculous in origin.

Why do you say so ?

This is a case of pleural effusion (fluid collected within pleural cavity) because there are presence of:

(A) SYMPTOMS (from the history) :

- Evening rise of temperature for 1 month.
- Dry cough with left-sided chest pain for 20 days.
- Heaviness on left side of chest for last 15 days.
- Respiratory distress for last 3 days.

(B) SIGNS :

1. **General survey :**

- Decubitus—Prefers to lie on left lateral decubitus.
- Respiration—26/min, abdomino-thoracic in type and the rhythm is regular. There is less movement on left side of chest.
- Lymph nodes—Not palpable (rarely, few matted non-tender or slightly tender cervical lymph nodes may be palpable in tuberculosis).
- Pulse—90/min, regular; mention other points.
- Temperature—100.8°F (oral) at present.
- Emaciation.
- Nicotine staining of the fingers (e.g., pleural effusion developing from bronchogenic carcinoma)—absent.

2. **Inspection :**

- Upper respiratory tract is within normal limit.
- Increased respiratory rate of 26/min.
- Fullness of intercostal spaces on left side of chest.
- Diminished movement of chest on left side.
- Apical impulse could not be visualised.
- Absence of intercostal suction.

3. **Palpation :**

- Diminished movement on left side of chest and the expansion is only $1\frac{1}{2}$ " as measured by the tape.
- Trachea is shifted to the right side and apex beat could not be localised (i.e., impalpable) (if apex beat can be localised in a left-sided pleural effusion, it will be situated somewhere near the sternum, i.e., towards the right side).
- Vocal fremitus is diminished on left side.
- Absence of tenderness.

- e) Friction fremitus—absent.
- f) Normal palpatory findings on right chest.

4. Percussion :

Stony dullness on the whole of left side of chest which does not alter with change of posture, i.e., shifting dullness is absent. Traube's space is dull on percussion. Tenderness is absent while percussing the chest. Normal resonant note on right side of chest.

5. Auscultation :

- a) Diminished vesicular breath sound on left side (or absent breath sound). Normal vesicular breath sound on right side.
- b) Vocal resonance—Diminished on left side; normal vocal resonance on right side.
- c) Adventitious sounds —Crepitations, rhonchi or pleural rub is absent both in left and right side of chest.

* Sometimes, there is tubular breath sound, bronchophony and whispering pectoriloquy (with increased vocal fremitus), and aegophony present at the upper border of pleural effusion.

* * Vocal fremitus, percussion and auscultation are carried out in the following way :

1. Anterior chest—Along MCL (supraclavicular [apex], infraclavicular, mammary and inframammary area).
2. Lateral chest—Along MAL (axillary and infra-axillary area).
3. Back —
 - (i) Upper part—Suprascapular area,
 - (ii) Middle part—Interscapular area,
 - (iii) Lower part—Infrascapular area (along scapular line)

*** According to few clinicians, 'slow' shifting dullness may be present in pleural effusion though it is difficult to demonstrate (not accepted by all).

**** The stony dullness of 'right'-sided pleural effusion is continuous with liver dullness while in 'left'-sided pleural effusion, the dullness is continuous with cardiac dullness with loss of Traube's space tympanicity.

What is your case ?

Build up the summary from the symptoms and signs, and try to say systematically as described above.

Why it is not a case of pneumothorax ?

Presence of stony dullness on percussion rules out pneumothorax (pneumothorax is tympanitic on percussion) and establishes the diagnosis of pleural effusion.

Why it is not a case of hydropneumothorax ?

As there is absence of :

1. Shifting dullness,
2. Succussion splash, and
3. Classical horizontal fluid level.

Important points in general survey in respiratory system :

1. **Decubitus**—May be propped-up; orthopnoea.
2. **Nutrition**—Emaciation in tuberculosis, bronchogenic carcinoma, diabetes mellitus and AIDS (DM and AIDS are often associated with pulmonary tuberculosis).
3. **Fades** — Moon face due to SVC syndrome, sarcoidosis (lupus pernio).
4. **Anaemia**—Tuberculosis, bronchogenic carcinoma, massive haemoptysis.
5. **Jaundice**—Bronchogenic carcinoma metastasizing in liver, antituberculosis drug-induced hepatitis, disseminated tuberculosis, pulmonary thromboembolism.
6. **Cyanosis**—Acute severe asthma, tension pneumothorax, lobar pneumonia etc.
- 7 **Clubbing**— Bronchogenic carcinoma, bronchiectasis, lung abscess, empyema thoracis etc.
8. **Pulse**—Pulsus paradoxus in acute severe asthma.
9. **Lymph nodes**—Specially the supraclavicular glands, scalene nodes, Virchow's gland and glands in the posterior triangle of neck.
10. **Respiration**—Tachypnoea, orthopnoea, hyperpnoea etc.
 11. **Temperature**—For any infective aetiology like tuberculosis, lung abscess, pneumonia etc.
 12. Any obvious deformity —Gibbus (due to caries spine), kyphoscoliosis, different chest deformities.
13. **Oedema**—From chronic cor pulmonale.

14. Eye—Horner's syndrome (bronchogenic carcinoma), subconjunctival haemorrhage (chronic cough, SVC syndrome), chemosis of conjunctiva (SVC syndrome), polycythemia (COPD), phlyctenular keratoconjunctivitis (primary tuberculosis), choroid tubercle in retina (miliary tuberculosis), colour blindness (ethambutol toxicity), papilloedema (COPD and SVC syndrome).
15. Hands—Nicotine staining, clubbing, cyanosis, wasting of small muscles of one hand (thoracic inlet syndrome), warm and moist hand (type II respiratory failure) and flapping tremor.
16. Skin—Erythema nodosum, scars and sinuses in neck, scrofuloderma.

What is pleural effusion ?

Collection of serous fluid within the pleural cavity (in between visceral and parietal layers of pleura) is *pleural effusion*. Only when the character of the fluid is transudate, it is known as *hydrothorax*.

Actually, the collection of fluid (resulting from increased hydrostatic pressure or decreased plasma oncotic pressure) occurring with a 'normal pleura' is ultrafiltration of plasma (transudate) in **hydrothorax**; it is usually bilateral. The effusion due to 'pleural diseases' more nearly resemble plasma (exudate) which results from increased capillary permeability; **pleural effusion** is commonly unilateral but may be bilateral.

Causes of pleural effusion :

(A) Exudative (pleural effusion) :

- a) Pneumonia (bacterial and viral)—possibly the commonest cause worldwide.
- b) Tuberculosis (possibly the commonest cause in India).
- c) Bronchogenic carcinoma.
- d) Pulmonary infarction (may be transudative in character).
- e) Collagen vascular diseases like SLE, rheumatoid arthritis.
- f) Pleural mesothelioma.
- g) Lymphoma and leukaemias.
- h) Meig's syndrome (many authorities believe it to be transudative in nature).
- i) Pancreatitis, subphrenic abscess, liver abscess, oesophageal perforation, drug-induced,
- j) Asbestosis-associated pleural effusion (benign).
- k) Yellow nail syndrome (rare; due to lymphoedema).
- l) Post-myocardial infarction syndrome (rare).

(B) Transudative (hydrothorax) :

- a) Congestive cardiac failure (CCF).
- b) Nephrotic syndrome.
- c) Cirrhosis of liver.
- d) Hypoproteinaemia with severe anaemia.
- e) Constrictive pericarditis, pericardial effusion.
- f) Myxoedema.
- g) SVC syndrome.
- h) Peritoneal dialysis.

a) b) c) and d) under (A) are most common causes of pleural effusion.

How to diagnose hydrothorax ?

1. Affection is usually bilateral.
2. Presence of anasarca.
3. Evidence of CCF, cirrhosis of liver, nephrotic syndrome or severe anaemia with malnutrition.
4. Absence of signs of inflammation (i.e., no H/O fever, local tenderness etc).
5. Transudative nature of pleural fluid on aspiration.

In patients with hydrothorax, always look for collection of fluid in other serous sacs. So, one should be careful in detecting oedema, pericardial effusion, ascites, hydrocele etc.

Haemorrhagic pleural effusion (haemothorax) :

1. Neoplasm—primary or secondary, pleural mesothelioma.
2. Chest trauma (during paracentesis or placement of central line).
3. Tuberculous effusion.
4. Bleeding diathesis.
5. Lymphoma and leukaemias.
6. Anticoagulant therapy.

7. Pulmonary infarction.
8. Acute haemorrhagic pancreatitis.

* Pleural effusion resulting from mesothelioma usually does not shift the mediastinum as mesothelioma encases the lung on the affected side and is commonly related to asbestos exposure.

Chylous (milky) pleural effusion :

It results from leakage of chyle into the pleural space and is due to,

- | | |
|------------------|---|
| 1. Tuberculosis. | 5. Myxoedema (gold paint effusion). |
| 2. Malignancy. | 6. Trauma to the chest wall. |
| 3. Lymphoma. | 7. Congenital absence of thoracic duct. |
| 4. Filariasis. | 8. Yellow nail syndrome. |

Bilateral pleural effusion :

- | | |
|-----------------------------|---|
| 1. Rheumatoid arthritis. | 4. Bilateral tuberculous effusion (rare). |
| 2. SLE. | 5. Pulmonary infarction. |
| 3. Lymphoma and leukaemias. | 6. Pleural metastases. |

* Remember, hydrothorax is usually bilateral.

Predominant right-sided pleural collection :

- | | |
|--|------------------------------------|
| 1. Rupture of amoebic liver | 4. Meig's syndrome (fibroma of the |
| abscess into pleural cavity. ovary with ascites and pleural effusion). | |
| 2. Cirrhosis of liver. | 5. Right-sided subphrenic abscess. |
| 3. Congestive cardiac failure. | |

Predominant left-sided pleural collection :

1. Acute pancreatitis.
2. Oesophageal rupture.
3. Dressler's syndrome (post-myocardial infarction syndrome).
4. Left-sided subphrenic abscess.
5. Aortic dissection.

Recurrent pleural effusion (reappearance after thoracentesis) :

- | | |
|--------------------------------|--------------------------------|
| 1. Bronchogenic carcinoma. | 4. Collagen vascular diseases. |
| 2. Pulmonary tuberculosis. | 5. Pleural mesothelioma. |
| 3. Congestive cardiac failure. | 6. Lymphoma. |

Drug-induced pleural disease and/or pleural effusion :

- | | |
|--------------------|-----------------------|
| 1. Nitrofurantoin. | 4. Dantrolene sodium. |
| 2. Methysergide. | 5. Bromocriptine. |
| 3. Amiodarone. | 6. Procarbazine. |

* The pleural fluid is eosinophilic in drug-induced pleural effusion.

Bedside diagnosis of malignant pleural effusion :

Elderly patient with H/O smoking, nicotine stain in hand, emaciation, clubbing, palpable scalene nodes and may be with radiation mark over anterior chest. There is rapid and repeated collection of pleural fluid after thoracentesis, and the fluid may be haemorrhagic.

Causes of empyema thoracis (purulent effusion) :

Collection of pus within the pleural cavity is known as empyema thoracis. The causes are,

1. Rupture of tuberculous cavity or post-pneumonic.
2. Secondary infection of haemothorax.
3. Rupture of subphrenic abscess; perforation of oesophagus.
4. Rupture of lung abscess (mostly staphylococcal—thin walled and multiple).
5. Septicaemia or pyaemia.
6. Traumatic (penetrating wounds of chest, after chest aspiration or liver biopsy).

Bedside diagnosis of empyema thoracis (purulent effusion) :

1. Patient looks toxic and prostrated; loss of weight.
2. Hectic rise of temperature with rigors and sweating.

3. Tachycardia as well as tachypnoea.
4. Development of clubbing.
5. Intercostal tenderness (patient winces with pain) as well as fullness.
6. Skin is red, oedematous and glossy overlying empyema.
7. Signs of pleural effusion.
8. Rarely, a swelling in the chest wall is seen which gives impulse on coughing—'empyema necessitatis' (i.e., pus collected under the skin of the chest wall due to rupture of purulent material from the pleural cavity, which communicates with the empyema thoracis).

What is meant by 'stony dullness' on percussion ?

1. Dull note elicited by percussion, PLUS
2. Feeling of increased resistance by the percussing finger.

Bronchial breath sound above the fluid level in pleural effusion—mechanism behind :

It is said that air conducted through the relaxed or collapsed lung (compression collapse) produces tubular breath sound (so, there is bronchophony and whispering percutiloquy). Conductivity of the sound through collapsed (airless) lung is increased if the bronchus remains patent.

Classical findings at the upper border of moderate pleural effusion :

1. Actually there is horizontal upper border of fluid level. Ellis's S-shaped curve (i.e., highest level of dullness is in axilla in comparison to anterior and posterior wall, and thus assuming the shape of the letter 'S') is not demonstrated clinically. It is said that the upper border of fluid level sweeps towards the axilla due to capillary action, and the highest point of fluid in axilla is just a radiological observation.
2. Skodaic resonance (after Josef Skoda) on percussion —It is observed just above the upper border of moderate effusion and the note (hyperresonant) mimics the percussion over an empty wooden box (boxy note). It is due to compensatory emphysema.
3. Bronchial breath sound (tubular)—May be audible.
4. Pleural rub, rarely.
5. Aegophony.

Who keeps the trachea in the midline ?

The equally negative intrapleural pressure on both sides.

Is it possible to have trachea shifted towards the side of pleural effusion ?

It is possible in a case of pleural effusion (malignant) developing from ipsilateral bronchogenic carcinoma. If we go through the basic physiology, it is very easy to understand.

(A) In absorption collapse (due to endobronchial growth, i.e., bronchogenic carcinoma or foreign body impaction) :

1. Bronchus—Not patent (obstructed),
2. Intrapleural pressure—Remains negative, and
3. Trachea—Shifted to the same side. It is the more negative intrapleural pressure which draws the trachea to same side.

(B) In compression collapse (due to pleural effusion, pneumothorax or hydropneumothorax) :

1. Bronchus—Patent,
2. Intrapleural pressure—Positive, and
3. Trachea—Pushed to the opposite side due to positive intrapleural pressure.

Now, if we deal with a case of bronchogenic carcinoma with malignant pleural effusion, the tussle between (A) and (B) will determine the position of trachea. If (B) plays more, trachea will go to the opposite side and if (A) plays more, **trachea will be shifted towards the same side**. If (A) and (B) play equally, trachea will remain in the midline.

So, if the pulling effect on trachea by absorption collapse (i.e., bronchogenic carcinoma) overcomes the pushing effect on trachea by compression collapse (i.e., pleural effusion), trachea remains on the same side of effusion.

Red flag signs in pleural effusion :

1. Trachea shifted to the side of effusion.
2. Developing into empyema thoracis.

Pleural effusion present but trachea remains in the midline :

The possibilities are :

1. Mild pleural effusion (only the apical impulse is shifted to opposite side).
2. Malignant pleural effusion secondary to bronchogenic carcinoma (as described above).
3. Loculated or encysted pleural effusion (both the apical impulse and trachea may not shift).
4. Bilateral pleural effusion (if the amount of collection is more or less equal).
5. Pleural effusion associated with mesothelioma of pleura.
6. Coincidental association of pleural effusion with ipsilateral apical fibrosis.

Observation on inspection of 'back' in respiratory system :

Inspection of back (in respiratory system or CVS) **is always done in standing position** (if the condition of the patient permits) to avoid undue obliquity. Go to the back of the patient and look for :

1. Kyphosis (look from sides in profile).
2. Scoliosis (may be responsible for mediastinal shifting)
3. Drooping of the shoulder (signifies apical fibrosis or collapse, or trapezius paralysis).
4. Winging of the scapula (long thoracic nerve palsy or spinal deformity—Prominent medial border of scapula).
5. Whether the inferior angle of scapula on both sides are at the same level or not.
6. Interscapular area (spino-scapular distance)—Comparison between two sides of chest.
7. Intercostal suction.
8. Gibbus (acute angulation in the spine due to collapsed vertebra which is commonly developing from caries spine, and sometimes as a result of metastasis, or trauma).
9. Ankylosing spondylitis (stiff and immobile spine)—May produce restrictive lung disease.
10. Crowding of ribs (both in the front as well as the back).
11. Skin condition—Scar, sinus, herpes zoster, local oedema, venous prominence, arterial pulsation (Suzman's sign), pigmentation, deformity after thoracic operation, e.g., thoracoplasty etc.
12. Movement—Whether both the sides are moving simultaneously and symmetrically, or not.

Importance of past history in pleural effusion :

Any H/O :

1. Trauma.
2. Tuberculosis.
3. Asbestosis (mining, textile workers,, pipe fitters)—So, occupational history is important.
4. Diabetes mellitus.
5. Smoking (for malignant pleural effusion).
6. Skin rash, joint pain, neck swelling (lymphadenopathy)—For collagen vascular diseases, lymphoma etc.
7. Swelling all over the body (anasarca with hydrothorax).
8. Haemoptysis (tuberculosis, bronchogenic carcinoma etc).
9. Same type of illness in significant past (recurrence) or in the family (e.g., tuberculosis).

Differential diagnosis (D/D) of your case :

(A) Pneumonic consolidation—

1. Acute onset with high fever, rusty sputum and chest pain.
2. Pulse : respiration = 2 : 1 or 3 : 1 (normal ratio is 4 : 1).
3. Trachea and apex beat—**Normal** in position (i.e., no shifting).
4. Percussion—**'Woody dullness'**.
5. Auscultation—Tubular breath sound, bronchophony, whispering pectoriloquy, aegophony, crepitations and pleural rub may be present.

(B) Thickened pleura* —

1. Long history.
2. Crowding of ribs with drooping of the shoulder. Depression of intercostal spaces with reduced movement on the affected side.
3. Trachea and apex beat may be shifted towards the diseased side (in fibrothorax).
4. Dull note on percussion but **never stony dull**.

5. Diminished vesicular breath sound with diminished vocal resonance. Bronchial breath sound is never heard. Pleural rub may or may not be present.
6. Commonly an end result of tuberculosis, asbestosis or haemothorax.

* Often known as '**fibrothorax**' (diffuse pleural thickening when entire lung is surrounded by fibrotic pleura) and the clinical findings depend on degree of pleural thickening. There is blunting of costophrenic angle with tenting of diaphragm in chest X-ray.

(C) Fibrosis or collapse (absorption) of the lung—

1. Crowding of ribs, drooping of the shoulder and retraction of intercostal spaces: flat chest.
2. **Apex beat and trachea are shifted towards the affected side.**
3. Impaired resonance on percussion; never stony dull.
4. Diminished vesicular breath sound with occasional crepitations.

(D) Empyema thoracis— Already described.

(E) Bronchogenic carcinoma—

1. H/O cough, haemoptysis, chest pain and emaciation in an aged patient; smoker.
2. Superior mediastinal syndrome.
3. Cervical adenopathy (scalene group specially); clubbing.
4. **Usually no shifting of mediastinum** unless complicated by pleural effusion.
5. Localised rhonchi or crepitations.

(F) Pericardial effusion— Considered only in left-sided pleural effusion.

1. Trachea is in normal position (i.e., no mediastinal shift).
2. Tympanicity on Traube's space percussion—retained (lost in left-sided pleural effusion).
3. JVP—Engorged and may be pulsatile. Kussmaul's sign may be present.
4. Pulsus paradoxus.
5. Apex beat (if possible to localise) is medial to outer border of cardiac dullness.
6. Muffled heart sounds.

(G) Subphrenic abscess—

1. H/O recent abdominal operation or appendicitis etc.
2. Fever (hectic) with chill and rigor; pain in upper abdomen.
3. Rigidity in upper abdomen.
4. Absence of stony dullness in chest (on percussion).

[(H) Liver abscess— Considered only in right-sided pleural effusion.

1. H/O intestinal amoebiasis may or may not be present.
2. Patient looks toxic; fever with chill; shiny skin over the right lower chest.
3. Severe upper abdominal pain with right-sided intercostal tenderness present.
4. Upper border of liver dullness goes upwards.)

Confirmation of your diagnosis :

1. Chest X-ray — PA and lateral view, PLUS
2. Aspiration of pleural fluid (absolute proof).

Other findings on percussion in pleural effusion :

1. Area of dullness (triangular) on percussion against the vertebral column at the base of opposite lung (Grocco's triangle)—as a result of fluid forming a mediastinal bulge.
2. Loss of tympanicity of Traube's space (in left-sided pleural effusion).

Why there is dyspnoea in pleural effusion ?

Dyspnoea occurs if there is,

1. Massive collection and / or
2. Rapid collection.

Pleural effusion produces (compression) collapse of the lung with shifting of the mediastinum to opposite side which leads to reduction in the vital capacity.

Examination of CVS in respiratory diseases :

Search for dilated superficial veins over chest wall as well as engorged, non-pulsatile neck vein (SVC syndrome), pericardial effusion (as a complication of anasarca), and signs of RVH and RVF (chronic cor pulmonale).



Eliciting ankle jerk



Eliciting ankle jerk by special method (on kneeling patient)



Eliciting ankle clonus



Eliciting knee jerk



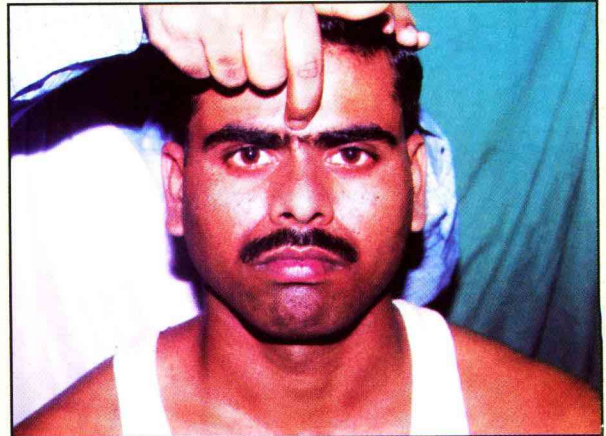
Eliciting knee jerk by special method with 'reinforcement'



Eliciting patellar clonus



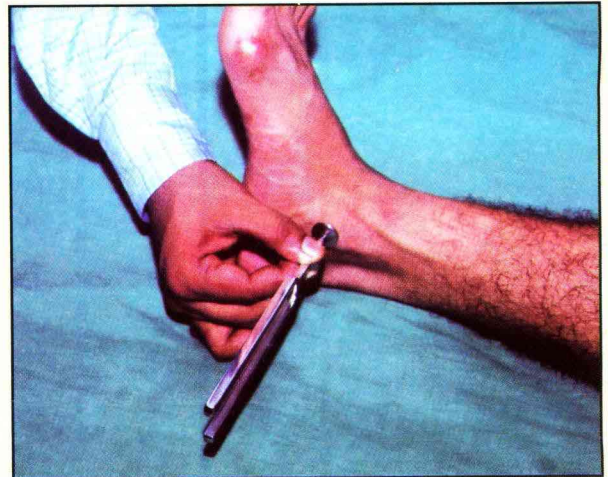
Testing **jaw jerk**



Eliciting **glabellar tap** standing behind the patient



Demonstration of **heel-knee test**



Testing **vibration sensation** at the ankle



Testing **joint-position sense**



Elicitation of **plantar response**

Examination of G. I system in respiratory diseases :

1. Halitosis (e.g., lung abscess).
2. Hepatosplenomegaly— Liver or spleen may be palpable due to the push effect of massive pleural effusion commonly rather than the cause of pleural effusion. Liver may be soft and tender in chronic cor pulmonale (liver of CCF). Splenomegaly is present in lymphoma, miliary tuberculosis, SLE and amyloidosis.
3. Ascites— In patients with hydrothorax (e.g., CCF), or disseminated tuberculosis.
4. Hydrocele or scrotal oedema— In patients with hydrothorax.
5. Abdominal lump— Pre- and paraaortic lymph nodes are palpable in lymphoma (may produce pleural effusion or SVC syndrome).
6. Hernial sites—Should be examined as protracted cough is common in COPD.

Examination of nervous system in respiratory diseases :

1. Neck rigidity / stiffness— If present, goes in favour of tuberculous meningitis or meningism due to pneumonia.
2. Higher function may be altered in SVC syndrome or respiratory failure.

* Horner's syndrome (bronchogenic carcinoma), myasthenic syndrome (Eaton-Lambert syndrome) and peripheral neuropathy may develop; the last two are non-metastatic complications of bronchogenic carcinoma.

Lymphadenopathy in a case of pleural effusion :

Think of :

1. Tuberculosis (specially in the posterior triangle of neck, supraclavicular fossa, axilla).
2. Lymphoma (generalised lymphadenopathy).
3. Bronchogenic carcinoma (scalene node, Virchow's gland).
4. Acute leukaemias (cervical, axillary).
5. SLE (generalised lymphadenopathy).

* Splenomegaly with pleural effusion is common in lymphoma, miliary tuberculosis, acute leukaemias, collagen vascular diseases (e.g., SLE) and cirrhosis of liver (hydrothorax).

What is 'phantom tumour' ?

Occasionally, effusion (transudate) forms between the lobes of lung (interlobar effusion) and produces a rounded opacity in chest X-ray, which mimics a solitary pulmonary nodule or mass lesion (sharply margined with bi convex appearance). The mass usually disappears with the resolution of effusion and this is why they are known as phantom tumour. Phantom tumour may be seen in congestive cardiac failure, and 'vanishes' after diuretic therapy.

Sub-pulmonic effusion and its diagnosis :

Here the effusion collects underneath the lung and the chest X-ray (PA view) gives an impression of elevated hemidiaphragm. To confirm the diagnosis, a chest X-ray in 'LATERAL DECUBITUS' position (affected side down) is taken which shows the pleural fluid layering out along the lateral chest wall.

Clinically, the left-sided sub-pulmonic effusion is diagnosed by loss of Traube's space tympanicity and the right-sided effusion is suspected in abnormal tidal percussion on right side.

How massive pleural effusion is diagnosed clinically ?

In massive pleural effusion, the dullness (on percussion) extends upto the 2nd ICS or above. Radiologically, it produces dense opacification of hemithorax with mediastinal shift. Usually it is designated as mild (< 500 ml), moderate (500-1500 ml) and massive (>1500 ml) according to the collected amount of fluid. Massive effusion commonly results from malignancy, CCF, tuberculosis, cirrhosis of liver and trauma.

Points in favour of tuberculous aetiology in your patient :

It is possibly the commonest cause of pleural effusion in India. The points in favour are :

1. H/O evening rise of temperature and cough for approximately 1 month.
2. Presence of anorexia, night sweats, malaise and loss of weight for last 2 months.
3. H/O contact.
4. Emaciation.

* Tuberculous effusion Is thought to be due primarily to a hypersensitivity reaction to tuberculous protein. Other hypersensitivity reactions of tuberculosis are erythema nodosum and phlyctenular keratoconjunctivitis.

Complications of pleural effusion :

1. Thickened pleura.
2. Empyema thoracis.
3. Collapse (compression) of the lung followed by fibrosis later on.
4. Spread of infection (tuberculosis) to pericardium or peritoneum.
5. Cardio-respiratory embarrassment as a result of massive pleural effusion.
6. Acute pulmonary oedema (rare)— As a complication of rapid pleural aspiration.
7. Hydropneumothorax— May develop after pleural aspiration.

How do you like to investigate a case of pleural effusion ?

1. Blood—TC, DC. ESR, sugar (PP), serum amylase, any abnormal cell (high ESR and mild lymphocytosis goes in favour of tuberculous aetiology).
2. Chest X-ray (PA view) —
 - a) Triangular homogeneous opacity with a curved upper border which is concave medially and upwards, and extends upto axilla.
 - b) Obliteration of costophrenic angle (earliest site of fluid collection).
 - c) Massive effusion produces unilateral homogeneous opacity.
 - d) Displacement of mediastinum to the opposite side.
 - e) LATERAL VIEW is done to differentiate it from lobar consolidation.
- * Encysted pleural effusion may not obliterate the costophrenic angle.
- ** A lateral decubitus film may be taken in a suspected patient of sub-pulmonic effusion.
- *** Repeatability of chest X-ray should be done in all cases after therapeutic paracentesis thoracis.
3. Aspiration of pleural fluid—

For diagnostic purpose, at least 50 ml fluid should be aspirated. It helps by,

 - a) Confirmation of diagnosis of pleural effusion.
 - b) Examination of the nature of fluid—Transudate or exudate [physical character (clear in hydrothorax, red in haemorrhagic effusion, milky white in chylous and purulent in empyema thoracis), biochemical (TLDH in exudate, adenosine deaminase activity in tuberculosis), and cytological (RBC, WBC, malignant cells) examination].
 - c) Smear examination—

Gram's stain, Ziehl-Neelsen stain, Leishman's stain for microfilaria or special stains for detection of malignant cells.
 - d) Culture of the aspirated fluid (bacteriological including culture of *M. tuberculosis* in Lowenstein-Jensen media). Recently introduced culture technique (Bactec system) gives result within 7 days in favour of tuberculous effusion.

A chest X-ray should be repeated after aspiration of pleural fluid to see the pulmonary parenchyma, specially in a suspected case of bronchogenic carcinoma. Sometimes, ultrasonography (USG) is done to differentiate an encysted pleural effusion from pleural tumour, and to localise effusion before aspiration or pleural biopsy.
 - e) Special tests—Pleural fluid adenosine deaminase activity (Tin tubercubsis), and PCR study (polymerase chain reaction to detect organisms including tubercle bacilli).
4. Sputum examination— It is done for 3 consecutive days.
 - a) For demonstration of AFB (surest test for tuberculosis) — Rarely positive.
 - b) To diagnose the aetiology of pneumonia (if effusion develops from pneumonia).
 - c) For malignant cells.
5. Mantoux test (not much informative)—
 - a) Positive in cases of tuberculosis.
 - b) May be negative in lymphoma.
6. Fine needle aspiration cytology (FNAC) or excision biopsy of lymph nodes (axillary, cervical or scalene node) —• May give a clue to the aetiology.
7. Imaging—Ultrasonography, CT scan or MRI scan detects fluid as small as 50 ml, and may be done if facilities are available.
8. Pleural biopsy (by Abram's, Cope's or trucut needle) — Specially in malignancy or tuberculosis.

9. Serological tests like rheumatoid factor, antinuclear factors, complement level etc.
10. Bronchoscopy and biopsy (in a suspected case of bronchogenic carcinoma).
11. Thoracoscopy and biopsy (pleura is inspected after inducing an artificial pneumothorax).

Minimal collection of pleural fluid for clinical detection :

At least 500 ml of fluid is necessary for clinical detection (minimally 300 ml fluid is required for radiological detection).

Possible causes of malignant pleural effusion :

1. Bronchogenic carcinoma (commonest).
2. Metastatic carcinoma (often from breast or thyroid).
3. Pleural mesothelioma.
4. Lymphoma.
5. Acute leukaemias.

* These are the causes of rapid and repeated accumulation of pleural fluid.

Indications of paracentesis thoracis (thoracentesis) :

1. Diagnostic - For physical character, biochemical, cytological and bacteriological study of pleural fluid to come to a definite diagnosis. At least, 50 ml fluid should be aspirated.
2. Therapeutic - If there is,
 - (i) Respiratory distress,
 - (ii) Massive collection,
 - (iii) Rapid collection of fluid, and
 - (iv) Suspected secondary infection of effusion.

Fluid should be aspirated slowly and as much as possible until it is harmful for the patient. Few clinicians advocate not to aspirate more than 1 litre of pleural fluid on the first occasion because of the risk of developing acute pulmonary oedema. The area of maximum dullness on percussion (usually in the mid-axillary line) should be the appropriate site for aspiration of fluid.

Complications of pleural aspiration or pleural tapping (thoracentesis) :

It is always done under strict aseptic condition with slow withdrawal of fluid. The complications are,

1. Pleural shock.
2. Empyema thoracis.
3. Hydropneumothorax.
4. Acute pulmonary oedema (non-cardiogenic; unilateral) - If fluid is removed very quickly.
5. Air embolism.
6. Cardio-respiratory embarrassment with circulatory collapse.
7. Subcutaneous emphysema.

Special characteristics of tuberculous effusion :

1. Possibly the commonest cause of pleural effusion in India and is usually associated with primary tuberculosis.
2. Straw-coloured or amber-coloured; EXUDATE in character (see below).
3. > 50% lymphocytes with paucity of eosinophils.
4. Cobweb coagulum formed in pleural fluid.
5. pH is towards the acidic side.
6. Adenosine deaminase activity (ADA) in the fluid is elevated (>40 IU/L).
7. Others : elevated level of interferon-gamma (IFN- γ > 140 pg/ml), presence of mycobacterial DNA (detected by PCR).
8. Paucity of AFB in pleural fluid. Yield :
 - a) Pleural fluid—AFB is found in 6-8% cases.
 - b) Ultracentrifused fluid—AFB is detected in 25-30% cases.
 - c) Pleural biopsy—AFB is detected in 80% cases.
9. Pleural fluid culture for M. tuberculosis is 45% sensitive.

* 'Quantiferon-TB Gold test' is a test which detects latent as well as active tuberculous disease. The test measures a component of cell-mediated immune reactivity to M. tuberculosis and is based on quantification of IFN- γ released from sensitized lymphocytes. The test requires a blood sample and the result is available within 24 hours. It is not affected by prior BCG vaccination.

Table 4 : Differentiation between transudate and exudate

Tests	Transudate	Exudate
(A) Routine :		
1. Appearance	Clear or light yellow. No froth on shaking	Straw-coloured, cloudy, purulent, milky, haemorrhagic. Froth formation on shaking
2. Protein	< 3.0g / 100ml	> 3.0g / 100 ml
3. LDH	Low (< 200 IU/L)	High (> 200 IU/L)
4. Pleural fluid LDH	< 2/3rd of normal upper limit of serum LDH	> 2/3rd of normal upper limit of serum LDH
5. Pleural fluid LDH/ serum LDH ratio	< 0.6	> 0.6
6. Pleural fluid protein / serum protein ratio	< 0.5	> 0.5
(B) Cytology :		
7. RBC	< 10000/mm ³	> 100000/mm ³
8. WBC	< 1000/mm ³	> 1000 mm ³ (> 50% lymphocytes in tuberculosis and neoplasm; >50% polymorphs in acute infections)
9. Mesothelial or malignant cells	Not found	May be seen in neoplasm
(C) Special :		
10. pH	> 7.3	< 7.3
11. Specific gravity	< 1016	> 1016
12. Glucose	Same as blood concentration	Low in infections and neoplasm (very low in rheumatoid arthritis)
13. Amylase	—	High in pancreatitis, oesophageal rupture and neoplasm
14. Microbiology	Negative	Positive

* No. 4, 5 and 6 are known as *Light's criteria* in pleural effusion.

** Some clinicians opine that SFAG [serum-fluid albumin gradient (i.e., serum albumin minus fluid albumin)] value may determine the nature of the fluid, i.e., SFAG > 1.1 is transudate, while <1.1 is exudate. The same calculation may be done by 'serum protein' minus 'fluid protein' and if >3.1, it is transudate and if <3.1, it is exudative in nature.

How to assess the movement of the chest ?

One has to assess whether both sides of the chest are moving simultaneously and symmetrically, or not. This is conventionally done at 3 places :

(A) FRONT-

Place both palms laterally on the lower rib cage in such a way that the thumbs touch each other in the middle anteriorly with a fold of skin in between the thumbs. Place the palms on the chest wall at the end of expiration. Now note the separation of thumbs and feel over the palms with inspiration. Usually this is done *at the level just below the nipples ('lower anterior chest')*.

Normal expansion of the chest in an adult is at least 5 cm (5—8 cm normally), and expansion of 2 cm or less is abnormal. In severe emphysema and ankylosing spondylitis, it is found to be 1 cm

or less.

N.B. : According to many clinicians, the respiratory movement of the '*upper*' anterior chest is determined by placing both palms side by side with thumbs touching the midline (terminal part of fingers will fall over clavicles). Now, note the elevation or lifting of palms, and not the separation of thumbs to assess the movement of the '*upper*' anterior chest.

(B) BACK -

- Place the palms vertically side by side in the interscapular region. Note the elevation or lifting of palms with inspiration.

- b) At the infrascapular region—Same manoeuvre, as done in the 'lower' anterior part of the chest. Note the separation of thumbs with inspiration.

(C) APEX-

Patient will sit, and standing behind the patient place two palms on the shoulder (over the clavicle) in such a way that thumbs touch each other in the nape of the neck at the level of the vertebra prominens. Note the elevation or lifting of palms with inspiration (not the separation of the thumbs). The movement of the apex may be examined from the front in a patient who is unable to sit (patient will lie down and palms will be placed over the clavicles from the front).

After the clinical assessment of movement of the chest, always measure the expansion (in the lower part of anterior chest wall and infrascapular region of back) with a measuring tape. Take the measurement both in inspiration and expiration.

* The side with diminished expansion is likely to be the pathological side.

** Upper and middle lobe expansion is best assessed from the front while lower lobe expansion is measured from the back.

Restriction of chest movement :

(A) Unilateral :

1. Pleurisy.
2. Chest trauma*.
3. Pleural effusion.
4. Pneumothorax.
5. Hydropneumothorax.
6. Collapse of the lung.
7. Fibrosis of the lung.
8. Thickened pleura.
9. Consolidation of the lung.
10. Empyema thoracis.

(B) Bilateral :

1. Emphysema.
2. Diffuse interstitial fibrosis.
3. Hydrothorax (bilateral).
4. Ankylosing spondylitis.
5. Sometimes in bronchial asthma.
6. Weak inspiratory muscles, e.g., myasthenia gravis, respiratory paralysis (e.g., G.B. syndrome).
7. Any extensive bilateral disease.
8. Obesity.

* Due to severe chest pain, fractured ribs, flail segment following trauma or previous thoracoplasty.

What do you mean by 'expansion' of the chest ?

Expansion and movement of the chest are not interchangeable terms e.g., in emphysema, chest may move but there is little expansion. 'Expansion' is the difference between inspiration and expiration, and is commonly measured by a tape.

Why there is aegophony above the level of effusion ?

It is due to interceptions of the relaxed lung as well as the fluid level, with the low-pitched component of the sound.

Why the upper margin goes towards axilla in chest X-ray ?

This is a radiological illusion. A horizontal section of hemithorax at the level of the upper margin of fluid shows that there is same amount of fluid present anteriorly, posteriorly and laterally. But it is a fact that X-ray beam traverse more fluid laterally than they do centrally because of the peculiar shape of hemithorax. So we see the curved upper margin going towards axilla in X-ray picture.

Some clinicians opine that it is due to capillary suction between two pleural layers, which draws the fluid up.

How will you manage a case of pleural effusion ?

1. Rest in bed with good nutritious diet.
2. NSAID - To relieve chest pain.
3. Aspiration of pleural fluid if the patient suffers from respiratory distress or if there is rapid collection.
4. Chest physiotherapy should be encouraged to help in the expansion of lower chest.
5. According to aetiology :
 - a) Tuberculous -
 - (i) Antituberculosis chemotherapy.
 - (ii) Sometimes, corticosteroid is added to prevent adhesion (20-40 mg daily for 6-12 weeks with gradual tapering).
 - b) Malignancy-
 - (i) Repeated aspiration of pleural fluid.

- (ii) Pleurodesis (obliteration of the pleural space by adhesion) is done by tetracycline, bleomycin, mustine hydrochloride, talcom powder, Corynebacterium parvum etc. Alternatively, thoracotomy with pleurectomy or pleural abrasion may be tried.
- (iii) Treatment of bronchogenic carcinoma or lymphoma is done accordingly,
- c) Hydrothorax- Treatment of CCF or cirrhosis of liver is done accordingly.

CONCLUSION :

1. Always look for puncture mark, benzene stain or cotton seal in the lower chest (indication of pleural aspiration).
2. Shifting of mediastinum to the opposite side, stony dullness on percussion, and diminished (or absent) vesicular breath sound and vocal resonance on the affected side are **cardinal points for diagnosis**.
3. Examine for shifting dullness in the chest to exclude hydropneumothorax in all cases of pleural effusion.
4. Never forget to write measurement of the chest in two phases, i.e., both in full inspiration and in full expiration.
5. Always examine the upper respiratory tract (nose, nasal septum, air sinuses, pharynx) at the beginning of clinical examination of respiratory system,
6. **Do not forget to examine the breast, testes and spine in a patient of pleural effusion.**

Case 9

PNEUMOTHORAX

What is your diagnosis ?

It is a case of right-sided spontaneous pneumothorax of closed type, probably of tuberculous aetiology.

Why do you say so ?

It is a case of pneumothorax (air collected within pleural cavity) because there are presence of :

(A) SYMPTOMS (from the history)—

- a) Respiratory distress for 3 days,
- b) Pain in the right chest for 3 days, and
- c) Rise of temperature for 2 months.

* *Breathlessness and pain chest started suddenly.*

(B) SIGNS—

1. General survey :

- a) Decubitus—Propped-up position at present. The patient prefers to lie on right lateral decubitus and becomes breathless on left lateral decubitus.
- b) Respiration—30/min, abdomino-thoracic in type and regular in rhythm. There is less movement on right side of chest.
- c) Lymph nodes—Not palpable.
- d) Cyanosis—Absent at present.
- e) Pulse—84/minute and regular; mention other points.
- f) Temperature—100°F (oral) at present.

2. Inspection :

- a) Upper respiratory tract is within normal limit.
- b) Increased respiratory rate of 30/min.
- c) Intercostal spaces are full on right side.
- d) No intercostal suction; accessory muscles of respiration are not working at present.
- e) Diminished movement on right side of chest.
- f) Apical impulse is seen on left side of chest, below the left nipple and placed more outward than normal.

3. Palpation :

- a) Diminished movement on right chest. Expansion of chest is 1½" as measured by the tape.

- b) Trachea is shifted towards left side. Apex beat is palpable at left 5th ICS, 1½" outside the left MCL.
- c) Vocal fremitus is diminished (or absent) on right chest. Normal vocal fremitus on left side.
- d) Absence of local tenderness all over in the chest.

Percussion :

- a) Tympanitic note on all over the right chest.
- b) Obliteration (reduction) of liver dullness, and the upper border of dullness is present on right 7th ICS in the MCL.
- c) Shifting dullness—Absent.
- d) Normal resonant note on percussion over left side of the chest.

Auscultation :

- a) Breath sound is diminished vesicular (or absent) on right chest.
- b) Vocal resonance is diminished (or absent) on right chest.
- c) Abventitious sounds — Neither rhonchi nor crepitations are heard. Pleural rub is absent.
- d) Coin sound — Present on right chest.

Auscultatory findings on the left side of chest are normal.

* Pyrexia may be absent in pneumothorax; *chest pain and dyspnoea are two chief complaints.*

** *The chest in pneumothorax is known as 'tympanicity with silence'* (i.e., tympanitic note on percussion with diminished or absent breath sound).

*** A small pneumothorax may be asymptomatic without any abnormal physical signs in chest.

What is your case ?

Build up the summary from the symptoms and signs mentioned above.

Describe the dyspnoea (H/O present illness) in your patient :

The dyspnoea is due to collapse (compression) of the lung. It is,

1. Acute (sudden) in onset which compelled the patient to attend hospital immediately.
2. Sense of something being torn inside the chest, at the beginning of symptoms.
3. Not associated with wheeze (may be associated with wheeze in a COPD patient).
4. Dyspnoea at rest.
5. Not relieved by drugs, rest, squatting etc; minimally relieved by adopting sitting posture or by right lateral decubitus position.
6. Associated with chest pain.
7. Non-progressive.
8. No H/O PND.
9. Not associated with 'shock and collapse' at the onset.
10. Cough aggravated the breathlessness.

* For gradation of 'respiratory' dyspnoea, see page 96.

Describe the chest pain (H/O present illness) in your patient :

1. It is sharp, and stabbing or tearing in nature; acute in onset and excruciating.
2. Present on the right side of the chest.
3. Increases by coughing, laughing, sneezing, jolting and inspiration.
4. Associated with dry cough.
5. The patient is forced to take shallow breathing and suppression of coughing.
6. Not relieved by any drug or by any other measure; forced the patient to attend hospital.
7. Without any radiation.
8. In all respect, it is a pleuritic pain.

Importance of past history in pneumothorax :

History of ;

1. Respiratory distress (COPD may be an aetiology of pneumothorax).
2. Tuberculosis.
3. Haemoptysis.
4. Trauma.

5. Prolonged recumbency with calf pain (indicates pulmonary thromboembolism—as D/D of dyspnoea of sudden onset).
6. Similar episode (recurrent pneumothorax) in the past.
7. Chest pain (IHD) — Indicates acute myocardial infarction (D/D of dyspnoea and pain chest).
8. Systemic hypertension (LVF may be a D/D of acute onset dyspnoea).

Other possible auscultatory findings in pneumothorax :

1. Amphoric breath sound may be heard on the affected side (open type).
2. Pneumothorax click—Sharp clicking sound synchronous with the heart beat may be heard in left sided 'shallow' or small pneumothorax.
3. Hamman's sign (in case of pneumomediastinum)— Read the section on 'Pericardial rub'.

How to examine for subcutaneous emphysema ?

It is the presence of air in the subcutaneous soft tissue and is also known as 'surgical emphysema' (older term). When one gives light pressure after placing the fingers fanwise on the affected area, a peculiar spongy or crackling sensation (crepitus) is obtained. If a stethoscope is placed and pressed there, crepitations-like sound is heard. It may extend from face to the foot resulting in diffuse swelling of the part. The clinical associations are :

1. **Pneumothorax** (after intercostal tube drainage) or severe bronchial asthma (air may track into mediastinum resulting in mediastinal emphysema).
2. Rib fracture which penetrates the lung i.e., injury to the chest wall.
3. Fracture of paranasal air sinus.
4. Rupture of the oesophagus.
5. Laryngeal perforation (accidental).
6. Gas gangrene.

* Always search for subcutaneous emphysema in a case of pneumothorax having tube drainage.

Types of pneumothorax :

1. **ARTIFICIAL** - Artificially done to control tuberculosis by inducing collapse of the lung (not used now-a-days) or to differentiate radiologically among the lesion in pleura, lung and chest wall (diagnostic purpose).
2. **TRAUMATIC** - Chest trauma (stab injury), thoracic surgery, after pleural or lung biopsy, CVP recording through subclavian vein, fractured ribs, pericardiocentesis, bronchoscopy.
3. **SPONTANEOUS** -
 - a) Closed [mild (<20% of radiographic volume in chest X-ray), moderate (20-50%) and large (>50%) variety] - Usually develops due to rupture of emphysematous bulla or from rupture of subpleural tuberculous focus. The communication between pleura and lung closes by fibrin clot as well as collapse of the lung, and usually does not reopen. Dyspnoea is not very severe and infection of pleural space does not occur. There is presence of mediastinal shift.
 - b) Open - There is development of free communication between pleural space and bronchus (**bronchopleural fistula**). Air goes in and out of the pleural space with inspiration and expiration respectively. Expansion of collapsed lung is hampered and infection of the pleural space is a common complication (pyopneumothorax). Open type usually originates from the tuberculous cavity, lung abscess or apical subpleural bleb. Amphoric breath sound may be auscultated. The patient is dyspnoeic and there is presence of mediastinal shift. Penetrating wound of the chest also produces open type of pneumothorax.
 - c) Tension (valvular) - The pleuro-pulmonary communication acts as a check-valve which allows air to enter into the pleural cavity during inspiration, coughing, sneezing and straining but prevents escape of air during expiration. Very large amount of air may be trapped within the pleural cavity in this way. Dyspnoea is very severe, and mediastinal shift is obvious and progressive (thus, there is shock and collapse). Central cyanosis is present. **It is a medical emergency** and the patient should be tackled immediately.

* Clinical classification of pneumothorax : closed type, open type and tension type.

Pneumothorax in relation to atmospheric pressure :

1. Closed type—Intrapleural pressure < Atmospheric pressure.
2. Open type—Intrapleural pressure = Atmospheric pressure.
3. Valvular type—Intrapleural pressure > Several times of atmospheric pressure.

What is normal intrapleural pressure ?

Normal Intrapleural pressure is -2.5 to -6 mm of Hg. It is the negative intrapleural pressure, i.e., -2.5 mm of Hg. (subatmospheric) which keeps the lungs in apposition with the chest wall (at the end of expiration). At the height of inspiration, it becomes more negative, i.e., -6 mm of Hg.

Recent classification of spontaneous pneumothorax :

Spontaneous pneumothorax is one where air develops within the pleural space without antecedent trauma to the thorax. It is basically of two types :

- (A) **'Primary'** spontaneous pneumothorax :
 - a) Without underlying lung disease.
 - b) Usually due to rupture of apical subpleural blebs of 1-2 cm in diameter,
 - c) In tall, thin individuals between 20-40 years of age, and
 - d) Usually in smokers.
- (B) **'Secondary'** spontaneous pneumothorax :
 - a) With underlying lung disease (different aetiology are mentioned below),
 - b) Usually from COPD,
 - c) Most common in older persons, and
 - d) More dangerous than primary variety because of underlying lung disease.

Aetiology of pneumothorax :

1. Rupture of an apical subpleural bleb or bulla (usually in young patients).
2. Rupture of a subpleural tuberculous focus or tuberculous cavity.
3. Rupture of the pulmonary end of pleuro-pulmonary adhesion.
4. Rupture of subpleural emphysematous bullae (usually in elderly).
5. Staphylococcal lung abscess.
6. Penetrating or non-penetrating trauma to the chest.
7. Bronchogenic carcinoma.
8. Pulmonary infarction.
9. Rupture of 'Honeycomb lung' (bronchiectasis, scleroderma, histiocytosis X disease, tuberous sclerosis etc).
10. Bronchial asthma.
11. Whooping cough.
12. Cystic fibrosis.
13. Rupture of oesophageal carcinoma.
14. Rupture of a tension cyst in lung.
15. Artificial pneumothorax.
16. Catamenial pneumothorax—rare disorder in women over 25-30 years, occurring within 48 hours of onset of menstruation; probably due to pleural endometriosis; recurrent and usually right-sided, treated by OC pills to suppress ovulation, or danazole, surgical explorations and pleurodesis.
17. Caisson's disease.
18. ARDS.

* 1, 2, 3 and 4 are principle causes of pneumothorax. Aetiology No. 1 is usually congenital and probably the commonest cause worldwide.

Causes of recurrent (more than twice) spontaneous pneumothorax :

1. Patients with emphysematous bulla or apical subpleural bleb.
2. Cystic fibrosis.
3. Rupture of bronchogenic carcinoma or oesophageal carcinoma.
4. Patient with lung cysts.
5. Catamenial pneumothorax.
6. Honeycomb lung.
7. AIDS.
8. Ehlers-Danlos syndrome, Marfan's syndrome.

Causes of bronchopleural fistula :

1. Tuberculous cavity.
2. Lung abscess.
3. Necrotising pneumonia.
4. Following lung resection.
5. Chest trauma.
6. Barotrauma.
7. Empyema thoracis.

* Bronchopleural fistula is often used synonymously with open pneumothorax.

Percussion note over normal lung :

Lung is resonant on percussion while abdomen is tympanitic. When the air in a cavity of sufficient size is set into vibration, the character of the sound becomes tympanitic, e.g., in the case of percussion over stomach. On the other side, the lung alveoli are small pockets of air separated by numerous septa which dampen down the sound from tympanitic to resonant note.

Differentiation between different characters of percussion note :

It is described below from higher to lower note of resonance :

- | | |
|---|--------------------------------------|
| (A) Resonant note : | (13) Dull note : |
| 1. Tympanitic (abdomen, pneumothorax) of the lung). | 5. Woody dullness (consolidation) |
| 2. Hyperresonant (emphysema) | 6. Stony dullness (pleural effusion) |
| 3. Resonant (normal lung) | |
| 4. Impaired resonance (thickened pleura) | |

Tympanitic note on percussion :

- | | |
|--------------------------------------|--|
| 1. Pneumothorax. | 4. Subpleural lung cyst. |
| 2. Superficial, big, empty cavity. | 5. Sliding hiatal hernia (left side only). |
| 3. Over a large emphysematous bulla. | 6. Over Traube's space. |

* Remember, tympanitic note is elicited on percussion over any hollow viscus.

Hyperresonant note on percussion :

1. Emphysema.
2. Occasionally over lung cyst.

Impaired note (slightly diminished normal resonance) on percussion :

- | | |
|--------------------------|-------------------------------|
| 1. Thickened pleura. | 4. Over pulmonary neoplasm. |
| 2. Fibrosis of the lung. | 5. Early pleural effusion. |
| 3. Collapse of the lung. | 6. Consolidation of the lung. |

Dull note on percussion :

1. Consolidation (**woody dullness**).
2. Pleural effusion, empyema thoracis and over 'hydro' part of hydropneumothorax (**stony dullness**).

Causes of acute chest pain :

1. Ischaemic heart disease (IHD), i.e., angina pectoris or acute myocardial infarction.
2. Acute dry pleurisy due to any cause (e.g., tuberculosis, pneumonia).
3. Spontaneous pneumothorax.
4. Diffuse oesophageal spasm (may be food-related), gastro-oesophageal reflux disease.
5. Acute pulmonary thromboembolism.
6. Musculoskeletal pain : rib fracture (trauma, cough), secondary deposits in rib (malignancy, multiple myeloma), costochondritis (Tietze's syndrome), abscess or furuncle on the chest wall, myositis, fibrositis, intercostal myalgia (Bornholm disease — intercostal muscle involvement by Coxsackie B virus infection), thrombophlebitis of anterior thoracic vein (Mondor's disease), arthritis of shoulder joint or spine, intercostal neuralgias.
7. Acute dry pericarditis.
8. Acute tracheobronchitis, mediastinitis.
9. Aortic stenosis, hypertrophic cardiomyopathy.
10. Dissecting aneurysm.
11. Herpes zoster on the chest wall.

12. Anxiety states/psychogenic (cardiac neurosis).
13. **Abdominal disorders** e.g., hiatal hernia, acute cholecystitis, acute pancreatitis, perforated peptic ulcer, ruptured amoebic liver abscess, splenic flexure syndrome.

* Local tenderness is found in 6 and 11; 2 and 7 may produce tenderness.

Enumerate the causes of 'retrosternal' chest pain :

- | | |
|---|--|
| 1. Ischaemic heart disease. | 6. Mediastinal tumour or mediastinal emphysema |
| 2. Oesophagitis or diffuse oesophageal spasm. | 7. Aneurysm of the aorta. |
| 3. Acute dry pericarditis. | 8. Dissecting aneurysm. |
| 4. Acute mediastinitis. | 9. Tracheitis. |
| 5. Diaphragmatic hernia. | 10. Psychogenic. |

* Causes of chest pain remain within :

a) Retrosternal (central), b) Pleural (non-central; causes of pleurisy), and c) Musculoskeletal pain (non-central).

** **'Precordial catch'**—Sudden, momentary stabbing pain at cardiac apex experienced by normal subjects; physiological and does not indicate organic heart disease.

*** Sharp stabbing left 'submammary pain' associated with anxiety is known as cardiac neurosis or Da Costa's syndrome.

Dyspnoea of acute or sudden onset :

1. Cardiac asthma (cardiogenic pulmonary oedema).
2. Acute severe asthma (status asthmaticus).
3. Acute exacerbation of chronic obstructive pulmonary disease (COPD), or acute viral/bacterial bronchitis.
4. Acute laryngeal obstruction (child—foreign body, diphtheria, oedema of glottis; adult — foreign body like dentures, loose teeth, tetany, oedema of glottis etc.).
5. Spontaneous pneumothorax (tension type).
6. Anaphylaxis (oedema of glottis).
7. Acute pulmonary thromboembolism.
8. Rapidly accumulating massive pleural effusion.
9. Diabetic ketoacidosis.
10. Lobar pneumonia.
11. Cardiac tamponade.
12. ARDS.
13. Hysterical hyperventilation (dyspnoea is more at rest than on exertion).

* 1, 2, 4, 5, 6 and 7 are very important causes. Acute obstruction of upper airway with food is known as 'cafe coronary'.

**** Classification of dyspnoea according to time course of onset :**

- a) In seconds—4, 5, 6 and 7
- b) In hours—1, 2, 9, 10, 11 and 13
- c) In days—3, 8 and 12
- d) In weeks to months—Severe anaemia, pleural effusion, COPD, weakness of respiratory muscles e.g., myasthenia gravis.

Aetiology of chest pain with dyspnoea :

- | | |
|-------------------------------------|---|
| 1. Spontaneous pneumothorax. | *5. Dissection of aorta. |
| 2. Acute myocardial infarction. | 6. Acute dry pleurisy (specially from consolidation). |
| 3. Trauma to the chest wall. | 7. Psychogenic. |
| 4. Acute pulmonary thromboembolism. | |

* Usually (No. 5) from Marfan's syndrome, hypertension, coarctation of aorta and pregnancy.

Aetiology of chest pain with circulatory collapse or syncope :

- | | |
|-------------------------------------|--------------------------------------|
| 1. Acute myocardial infarction. | 5. Cardiac tamponade. |
| 2. Tension pneumothorax. | 6. Acute pancreatitis. |
| 3. Acute pulmonary thromboembolism. | 7. Upper gastro-intestinal bleeding. |
| 4. Dissection of aorta. | |

Different types of fremitus over chest :

Fremitus is synonymous with 'palpable vibrations'. The different types are :

1. Vocal fremitus.
2. Rhonchial fremitus (palpable rhonchi; rhonchi are better felt than crepitations).
3. Friction fremitus (palpable pleural rub).
4. Tactile fremitus :
 - a) Palpable coarse crepitations,
 - b) Palpable subcutaneous emphysema,
 - c) Palpable pericardial rub.

* Except the vocal fremitus, others are sometimes called as '**palpable accompaniments**'.

** **Vocal fremitus** is the palpation of laryngeal vibrations on the chest wall when the patient speaks some words repeatedly in a constant tone and voice (e.g., 'one, one' or 'ninety-nine, ninety-nine'). Place the palm of the hand (thenar and hypothenar eminences) or the ulnar border of right hand over the intercostal spaces on the chest, comparing the corresponding areas on both sides. Always use the same hand for examining both the sides (use right hand). Avoid the area of cardiac dullness on left side and place the hand a bit laterally. Start from above downwards in the front, sides and back of the chest. For changes (diminished or increased) in vocal fremitus, see page 92 as causes for changes in vocal fremitus are exactly the same as that of vocal resonance.

How cough is analysed at the bedside ?

1. Duration (days/months/years); acute < 3 weeks, chronic > 3 weeks.
2. Variability (daytime/nocturnal/morning).
3. Precipitating factors (dust/fumes/pollen/cold air; lying down-^ gastro-oesophageal reflux disease or CCF).
4. Sputum analysis (see below).
5. Types (dry/wet/bovine->see below).
6. Haemoptysis — present or not.
7. Associated symptoms (post-nasal drip, gastro-oesophageal reflux disease or occult asthma —> these are common causes of long-standing, undiagnosed cough); wheeze? seasonal?
8. Chest pain (pleuritis) or breathlessness (COPD) — present or not.
9. H/O drug intake—ACE-inhibitor, p-blocker.

Types of cough :

1. Dry (non-productive) — Acute dry pleurisy, acute tracheobronchitis, interstitial lung disease.
2. Wet (productive) — Bronchiectasis, lung abscess, resolution stage of lobar pneumonia.
3. Hacking (pharyngeal cough) — Heavy smokers.
4. Whooping — Found in whooping cough. There is rapid succession of dry coughs which gather speed gradually and end in a deep inspiration during which the characteristic 'whoop' (noise) is heard.
5. Brassy (with a metallic sound) — In carcinoma of larynx.
6. Bovine (laryngeal cough) — In recurrent laryngeal nerve palsy, which is commonly due to bronchogenic carcinoma (the explosive nature of cough is lost).
7. Spluttering cough In tracheo-oesophageal fistula (cough during swallowing).
8. Barking Harsh and loud cough; found in epiglottitis and hysteria.
9. Nocturnal — Chronic bronchitis, LVF, tropical eosinophilia, post-nasal drip, aspiration.
10. 'Croupy' cough — Laryngitis, specially in children.
11. Foetid cough In bronchiectasis and lung abscess, there is foul smelling expectoration.
12. Recurrent cough since childhood Cystic fibrosis, childhood asthma, congenital heart disease, cystic disease of lung, hypogammaglobulinaemia.
13. Cough with postural variation (postural cough)—Bronchiectasis, lung abscess, gastro-oesophageal reflux disease (GERD), bronchopleural fistula.

Profuse purulent sputum :

- | | |
|-----------------------------------|---|
| 1. Bronchiectasis. | 4. Empyema thoracis ruptured into bronchus. |
| 2. Lung abscess. | 5. Tuberculous cavity. |
| 3. Sometimes, chronic bronchitis. | 6. Cystic fibrosis, resolving pneumonia. |

* 1, 2 and 4 are causes of **profuse and offensive** sputum. Infection with anaerobic organisms and necrotising pneumonia also produce offensive sputum.

**** Profuse sputum** means approximately a teacupful (> 100 ml) of sputum production per day.

How sputum (expectoration or 'phlegm') is analysed clinically ?

1. Amount (profuse or not).
2. Character (serous, mucoid, purulent, mucopurulent).
3. Colour (yellow, green, black, rusty, pinkish or anchovy-sauce like).
4. Odour or taste (offensive or not).
5. Mixed with blood (haemoptysis) or not.
6. Sputum production influenced by change of posture (bronchiectasis, lung abscess) or not.

How to palpate the trachea ?

Normal position of trachea in a healthy person is **central** or slight deviation to the right side.

Steps :

1. Trachea should be palpated in standing (most preferable) or sitting position with arms placed symmetrically on two sides and chin held in the midline. **Trachea should not be examined in lying down position** unless the patient is very ill.
2. One may fix the head of the patient with his left hand. Standing in front of the patient (patient looking directly forward), place the right index finger on the cricoid cartilage (trachea starts from lower border of cricoid) and slide it down over the tracheal rings upto the suprasternal notch.
3. Now slide the index finger in the angle between the sternomastoid muscle and trachea on both sides (never poke the finger, otherwise the patient may start coughing). **The angle is narrowed and feels more resistant on palpation with deviation of trachea to that side.**

* It is better to have the patient's and examiner's head at the same level by proper adjustment (while sitting). Always exclude goitre (by inspection only) before palpating the trachea.

Shifting of trachea and apex beat :

(A) To same side (pull effect) :

1. Fibrosis of the lung.
2. Collapse of the lung (absorption collapse).
3. Grossly thickened pleura.
4. Pneumonectomy.

B) To opposite side (push effect) :

1. Pleural effusion.
2. Pneumothorax.
3. Hydropneumothorax.
4. Empyema thoracis.
5. Pyopneumothorax.
6. Haemothorax, haemopneumothorax.

Tracheal shifting is absent in consolidation, emphysema, ease, bronchitis, bronchiectasis and lung abscess.

bronchial asthma, interstitial lung dis-

**** 'Mediastinal shifting'** means shifting of trachea (superior mediastinum) PLUS shifting of apex beat (inferior mediastinum). Trachea is commonly shifted in diseases of the upper part of chest (like shifting of trachea to same side in apical fibrosis or collapse) and predominantly apex beat is shifted in involvement of the lower part of chest (like shifting of the apex beat to opposite side in mild to moderate pleural effusion though a massive pleural effusion shifts the trachea too).

***** Remember, mediastinum may be shifted in scoliosis.**

summarise, **causes of displacement of apex beat** are LVH, RVH, mediastinal shift (pleuro-pulmonary diseases), thoracic deformity (e.g., scoliosis) and dextrocardia.

How to differentiate dextrocardia from dextroversion ?

In dextrocardia (congenital), heart is present in right hemithorax with apex pointing to the right while trachea remains central in position. In dextroversion due to extracardiac causes (e.g., left-sided pleural effusion or pneumothorax, or right-sided fibrosis or collapse of the lung), both trachea and apex beat will be shifted to the right.

To **differentiate dextrocardia from dextroversion, always palpate the trachea.**

What is sternomastoid sign ?

It is the undue prominence of lower part of sternomastoid muscle due to deviation of trachea to that side. This is also known as **Trail's sign.**

Shifting of trachea without any lung pathology :

1. Severe scoliosis.
2. Enlarged lymph nodes on one side of neck.
3. Huge enlargement of one lobe of thyroid gland.

4. Aneurysm of the arch of aorta.
5. Any superior mediastinal growth.

Elicitation of 'coin sound' :

It is classically found in pneumothorax (and rarely, in the 'pneumo' part of hydropneumothorax).

Coin sound is mostly found in a **tense pneumothorax** (elicited by coin percussion) and also known as bell sound, bell tympany, bruit-de-airain or 'distal anvil sound'. The clinical examination goes like this :

1. The observer stands at the back. A metallic coin (one rupee coin preferably) is placed over the upper part of front of the affected chest and is percussed with a similar type of coin when the observer listens at the back (just diametrically opposite to the point of percussion) with the diaphragm of stethoscope. A high-pitched tympanitic bell-like metallic sound (ringing sound) is heard in a patient with tense pneumothorax. It is necessary to compare the sound with the healthy side (which produces a dull thud as the normal lung does not transmit the ringing metallic sound distinctly). The patient helps here by fixing one coin in the anterior chest wall with his left hand and also by beating it with another coin with his right hand.

Or,

2. An alternative method : the patient fixes the diaphragm of observer's stethoscope over the anterior chest wall (observer stands at the back of the patient and throws the stethoscope over the patient's shoulder) while the observer himself puts a coin in the back and strikes it with the second coin. In these methods, one coin is acting as pleximeter finger while the other is serving the purpose of percussing finger.

D/D you will consider in this case :

(A) Causes of unilateral tympanitic or hyperresonant note with diminished breath sound :

1. Large emphysematous bulla—Symptoms of chronic bronchitis may be present.
2. Lung cyst May be difficult to distinguish from pneumothorax (in contrast to pneumothorax, there is no mediastinal shifting and absence of collapsed lung at hilum in lung cyst).
3. Superficial, big, empty cavity (connected with a patent bronchus)—
 - a) Cavernous breath sound,
 - b) Post-tussive suction,
 - c) Post-tussive crepitations,
 - d) Cracked-pot sound on percussion.
4. Sliding hiatal hernia May have chest pain, dysphagia, heart burn, respiratory distress due to nocturnal regurgitation. The whole chest is not tympanitic. Breath sound is not totally absent in **the left side of chest.**

(B) Causes of chest pain with dyspnoea :

1. Acute myocardial infarction Occurs in middle aged or aged persons, retrosternal chest pain with radiation to left hand, drenching sweat and shock. Breathlessness with signs of heart failure may be found.
2. Acute pulmonary thromboembolism—Sudden onset of pain chest with dyspnoea, haemoptysis and circulatory collapse are present. Tachycardia, low BP, elevated JVP, right ventricular gallop rhythm, loud P₂ are not uncommon. Normal resonant note on percussion over the chest with paucity of signs on auscultation. H/O prolonged recumbency or signs of thrombophlebitis may
3. Trauma to the chest—H/O trauma, 'point of tenderness' may be detected. Normal resonant note on percussion. Typical findings of pneumothorax are lacking.
4. Acute dry pleurisy—Dyspnoea is not so common but chest pain is present. Pleural rub is audible and no sign of pneumothorax can be detected.
5. Dissection of aorta—Abrupt onset of tearing retrosternal chest pain and dyspnoea (mimicking acute myocardial infarction) with pain referred to the back. There is discrepancy between carotid pulses, difference of BP in two arms, arrhythmia, shock and features of acutely developing aortic incompetence. No sign of pneumothorax is present. The patient is commonly hypertensive or having Marfan's syndrome or in the third trimester of pregnancy.
6. Massive collapse of the lung—H/O aspiration of foreign body may be present. Mediastinal shifting is towards the side of collapse. Impaired resonance note is revealed on percussion. Breath sound is diminished vesicular or absent.

Complications of pneumothorax :

- | | |
|-----------------------|--|
| 1. Hydropneumothorax. | 5. Thickened pleura. |
| 2. Pyopneumothorax. | 6. Circular collapse; acute respiratory failure. |
| 3. Empyema thoracis. | 7. Atelectasis of lung. |
| 4. Haemopneumothorax. | 8. Surgical emphysema and pneumomediastinum. |

How do you like to investigate this case ?

1. Chest X-ray (PA view)—Done in **erect posture**. Sometimes, the X-ray is taken **after full expiration**, specially in a small pneumothorax. The radiological features are :
 - a) Increased radiolucency on right side of the chest with absence of bronchovascular markings in that area.
 - b) Collapsed right lung is seen as a homogeneous opacity with sharp outline (present paracardiac or paravertebrally).
 - c) Shifting of mediastinum towards the left side.
 - d) The left lung is showing mottled opacity in the apical area (signs of pulmonary tuberculosis) — May be normal.
 - e) Costophrenic angles are clear.

* Remember, in large lung cyst or large bulla (mimics pneumothorax in chest X-ray) there is no mediastinal shifting.

2. Blood—TC, DC, ESR (increased lymphocytes with high ESR point towards tuberculosis).
3. Mantoux test—Positive in tuberculous aetiology.
4. Sputum for detection of AFB is done for consecutive 3 days along with culture for M. tuberculosis.
5. Lung function tests—FEV¹, FVC, PEFR etc. (to detect underlying emphysema, COPD etc).

Management of pneumothorax :

- (A) Small pneumothorax (less symptoms)—The air gets absorbed spontaneously within few days and no treatment is required. The patients with underlying lung disease (COPD) can not tolerate even a small pneumothorax and in that case air must be removed. Small pneumothorax needs close observation with serial radiographic monitoring.
- (B) Large pneumothorax or tension pneumothorax—O₂ inhalation and propped-up position. A self-retaining catheter is inserted into the pleural cavity (in sitting position) in **the 2nd ICS along the MCL (preferable)**, or in the 4th or 5th ICS behind the anterior axillary fold (more comfortable for the patient and cosmetic for a female patient though not used routinely). The tube is now connected to a water-seal drainage system. If no bubbling is seen for 24 hours, or the patient is relieved of dyspnoea, or the chest X-ray shows complete re-expansion of lung, the tube is removed. The whole process takes approximately 3-4 days.
- (C) Recurrent spontaneous pneumothorax—Chemical pleurodesis is done by intrapleural instillation of kaolin, talcom powder, minocycline, 50% glucose solution, iodised oil etc. Alternatively, parietal pleurectomy or pleural abrasion may be tried.
- (D) Antituberculosis chemotherapy, whenever indicated. Sometimes, antibiotics are used to prevent secondary infection; antibiotics are always used in open type.
- (E) SurgeFy—
 - a) Thoracoscopy and cauterisation of the breach in the pleura, or excision of the bulla.
 - b) Pleurectomy (in recurrent type when pleurodesis fails).
 - c) Decortication and closure of bronchopleural fistula, lobectomy or thoracoplasty.

* Surgery is usually required for open type (cauterization, or open thoracotomy and direct closure of fistula).

** The rate of reabsorption of air is about 1.25% of the total radiographic volume per day and thus, a 50% pneumothorax (occupying 50% of hemithorax) will take 40 days to resolve totally.

Immediate management of tension pneumothorax :

- (A) O₂ inhalation and propped-up position.
- (B) Do not wait for the chest X-ray, and immediately insert a wide-bore needle (16 G) into the pleural space through the 2nd ICS at MCL (in sitting position). If large amount of gas escapes from the needle, the diagnosis is confirmed and the patient gets relieved. The needle should be left in place until an intercostal tube drainage with water-seal drainage system can be arranged at the earliest opportunity. If the tension pneumothorax is dealt in this way, circulatory collapse (cardio-respiratory embarrassment) can be prevented.
- (C) Treatment of shock and circulatory collapse, if present (tension pneumothorax⁺ mediastinal shift plus severe hypoxaemia → diminished venous return → inadequate cardiac output).

Water-seal drainage : whether working properly or not ?

The tube coming out from the pleural cavity should be placed under water. Exit-tube (for air) will always remain above the water level in the bottle. Look for bubbling of air in the water. The whole channel is patent and working if :

1. During each expiration or coughing, more bubbling occurs, and
2. During inspiration, water column ascends within the tube, which remains under water.

* The bottle should be kept below the level of thorax. If the water-seal drainage is required to continue for more than a week, the tube should be removed (always remove after clamping it) and a new tube is attached (the then may change the site of insertion in the chest).

Reasons behind failure of the lung to re-expand in due time :

1. The lung is trapped, or
2. A major bronchus is obstructed, or
3. Presence of bronchopleural fistula, or
4. Water-seal drainage is not working properly.

Will you advise any respiratory exercise ?

Yes. The patient is advised to inflate incentive spirometer, air pillows, balloons or football bladder, which will help in expansion of the collapsed lung. Before discharge from hospital, the patient is advised not to smoke, and must stop to fly or dive for 3 months.

Basic treatment strategy in pneumothorax :

1. Closed type—Observation, water-seal drainage if necessary.
2. Open type—Water-seal drainage system; treat hydropneumothorax or pyopneumothorax. Consult thoracic surgeon. Treat with proper antibiotic.
3. Tension type—Always tackle this medical emergency by water-seal drainage system.

N.B. : PLUS, Antituberculosis chemotherapy, if indicated.

Pneumothorax is seen in X-ray plate : will you treat ?

If the patient suffers from :

1. Dyspnoea, and/or
2. Cyanosis, and/or
3. Gross mediastinal shifting, and/or
4. Pneumothorax occupying > 50% of hemithorax, and/or
5. With underlying lung disease (e.g., COPD), and/or
6. Circulatory collapse

—Immediate treatment by intercostal tube drainage with underwater seal is necessary.

Describe the antituberculosis treatment :

It is the standard practice for patients with pulmonary and lymph node disease ;

(A) Short course chemotherapy **for 9 months** with three drugs regimen :

1. Rifampicin- 450 mg/day, if < 50 kg and aged; 600 mg/day, If > 50 kg.
2. INH (isoniazid)- 300 mg/day.
3. Ethambutol- 25 mg/kg/day.
or Pyrazinamide- 20-35 mg/kg/day (maximum 2.5 g).
or Inj. streptomycin (I.M) - 1 g/day, if > 45 kg ; and 0.75 g/day, if < 45 kg.

'Initial phase' for 2 months with 1+2+3, and 'Continuation phase' for 7 months with 1+2.

(B) Short course chemotherapy **for 6 months** with four drugs regimen :

1. Rifampicin- 450 mg/day, If < 50 kg and aged; 600 mg/day, if > 50 kg.
2. INH (isoniazid)- 300 mg/day.
3. Ethambutol- 25 mg/kg/day.
4. Pyrazinamide- 20-35 mg/kg/day (maximum 2.5 g).

'Initial phase' for 2 months with 1 + 2 + 3 + 4, and 'Continuation phase' for 4 months with 1+2.

N.B.: If ethambutol has to be continued after 2 months for any reason (like rifampicin toxicity etc), the dose will be 15 mg/kg/day. Bone tuberculosis should be treated for 9 months while tuberculous meningitis should receive treatment for 1 year. All the drugs should be taken In empty stomach and In single daily dose. Pyridoxine 10 mg/day may be added to prevent peripheral neuropathy induced by INH.

Corticosteroids in tuberculosis :

As steroids reduce inflammation and limit tissue damage, steroids are now recommended in patients with tuberculous affection of the following though opinion differs from clinician to clinician—

1. Pleural effusion.
2. Pericardial disease.



Nephrotic syndrome. Note the puffy and rounded face. Left eye is virtually closed by periorbital oedema due to posture-dependent nature of oedema



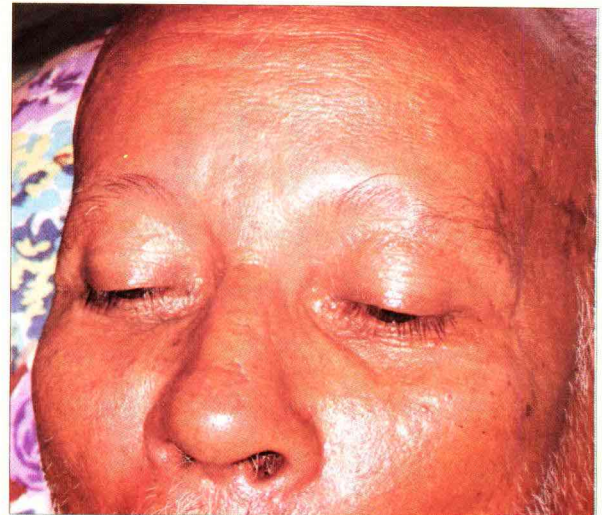
Scarring alopecia following bacterial folliculitis (inflammatory)



←
Chemotherapy-induced non-scarring alopecia



Hirsutism following prolonged corticosteroid therapy in bronchial asthma



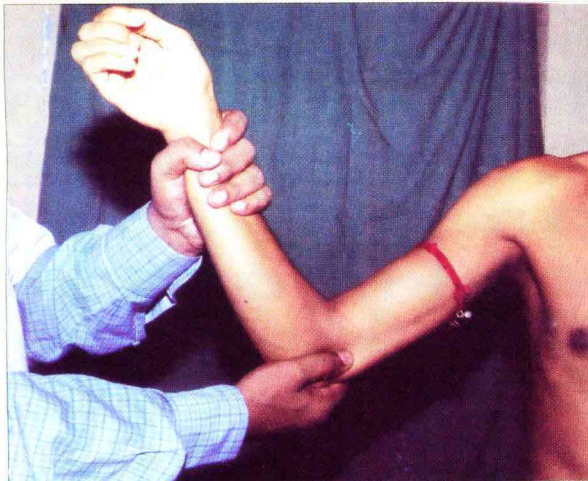
Madarosis – loss of lateral third of eyebrows in myxoedema



Detection of **clubbing** by bringing patient's fingers at eye level and looking tangentially



Palpation of **cervical glands** from behind using both hands



Palpation of **epitrochlear lymph nodes** of right side



Palpation of **axillary lymph nodes** of left side



Detection of **direction of venous blood flow** – a sitting patient with legs hanging from bed and by selecting a tributary-free vein while the patient is performing Valsalva manoeuvre to make the veins more prominent



Detection of **sternal tenderness** – pressing the upper part of body of sternum with ball of the right thumb

3. Meningeal disease or CNS tuberculosis.
4. Endobronchial disease (specially in children).
5. Extensive pulmonary disease.
6. Genitourinary tuberculosis.
7. Ophthalmic disease.
8. Hypersensitivity antituberculosis drug reactions.
9. Laryngeal or adrenal tuberculosis.
10. Phlyctenular keratoconjunctivitis.

Steroids should always be given under cover of antituberculosis chemotherapy.

Common side-effects of antituberculosis drugs :

Rifampicin—Nausea, vomiting, red-orange urine, flu syndrome, hepatitis, vasculitis, thrombocytopenia.

INH Peripheral neuropathy, hepatitis, lack of mental concentration, psychosis, convulsions.

Ethambutol—Optic neuritis, arthralgia, hypersensitivity (rare).

Pyrazinamide—Hepatitis, anorexia, nausea, vomiting, gout, fever, rash.

Streptomycin—Giddiness, loss of renal function, deafness.

PAS—Nausea, vomiting, goitre, blood dyscrasia, hypothyroid, hepatic damage.

Thiacetazone—Exfoliative dermatitis, gastro-intestinal reactions, hepatitis.

Cycloserine—Depression, psychosis, headache, somnolence, convulsions.

Ethionamide or prothionamide—Nausea, jaundice, neuropathy, metallic taste in mouth.

Kanamycin, capreomycin, viomycin—Giddiness, loss of renal function, deafness.

* **First five drugs are 'first line drugs' and others are 'second line drugs'. First four drugs are first line essential drugs while streptomycin, rifabutin and rifapentine are first line supplemental drugs.**

** Other drugs are—Rifabutin, rifapentine, quinolones (ciprofloxacin, sparfloxacin), macrolides (clarithromycin, azithromycin), amikacin, phenazines (clofazimine), linezolid, co-amoxycylav, different immunomodulators (cytokine immunotherapy).

What is DOTS ?

Directly Observed Treatment Short course (DOTS) chemotherapy is the backbone of revised National Tuberculosis Control Programme (RNTCP) in India. It is the administration of potent antituberculosis regimens to a patient having tuberculosis, in an intermittent manner, under direct supervision. As poor adherence to therapy is responsible for prolonged illness, relapse and resistance, supervised therapy 2-3 times per week definitely improves compliance. For DOTS, the patients are divided into 3 categories (I, II and III). Read DOTS in details from any standard text book.

Prognosis of pneumothorax :

It depends on :

1. Type of pneumothorax (closed, open or tension).
2. Amount of air present (indicates mediastinal shifting).
3. Underlying disease (emphysema, COPD).

* **It is important to remember that a large pneumothorax in a young individual may be less dangerous than a small pneumothorax in an aged patient with COPD.**

Conclusion :

1. Always look for puncture mark, benzene stain or cotton seal in the upper chest (anteriorly or axilla). If a tube or catheter is seen with water-seal drainage system, the diagnosis of pneumothorax is obvious (sometimes, a hydropneumothorax may be present with water-seal drainage).
2. Diminished or absent breath sound with tympanitic note on percussion on the affected side are **cardinal points for diagnosis** (there is loss of liver dullness in right-sided pneumothorax and loss of cardiac dullness in left-sided pneumothorax).
3. Search for shifting dullness in the chest to exclude hydropneumothorax in all cases of pneumothorax.
4. Remember, breath sound may be absent in both pneumothorax and pleural effusion, and in that case percussion note is the only guide (pneumothorax—tympanitic, pleural effusion—stony dullness) for diagnosis. Never forget that bronchial breath sound may be audible at the upper border of moderate pleural effusion as well as in case of open pneumothorax (bronchopleural fistula).

Case 10

HYDROPNEUMOTHORAX

What is your diagnosis ?

This is a patient of right-sided hydropneumothorax, probably of tuberculous in origin.

- * Hydropneumothorax is collection of both fluid (lower part) and air (upper part) in the pleura) cavity.

What are the cardinal features of hydropneumothorax ?

The hallmark of diagnosis in hydropneumothorax are 3 'S' :

1. Shifting dullness,
2. Succussion splash, and
3. Straight (horizontal) fluid level (upper limit of dullness is horizontal).

Is there any other feature present ?

1. In a sitting patient, the percussion note over air-containing upper part is tympanitic, and fluid-containing lower part is stony dull.
2. Tinkling sounds or metallic crepitations (specially after coughing) heard by stethoscope in upper chest, and
3. Positive coin sound in upper chest (rare).

Why do you say hydropneumothorax ?

One of the common aetiology of hydropneumothorax in our day to day practice is secondary infection of an open type of pneumothorax, or sympathetic collection of fluid in closed or tension pneumothorax. Thus, the patient will have symptoms like,

- | | |
|------------------------------------|----------------------------|
| a) Dyspnoea. | d) Cough |
| b) Pain chest. | e) Heaviness in the chest. |
| c) Splashing sound during jolting. | f) Pyrexia. |

The physical signs in hydropneumothorax are more or less similar to pneumothorax. While eliciting the signs mentioned above, one should be careful about (to differentiate from pleural effusion),

(A) During percussion -

- (i) There is horizontal fluid level (so called Ellis's S-shaped curve in pleural effusion, if any), and
- (iii) Shifting dullness (absent in pleural effusion).

(B) During auscultation, one should search for -

- (i) Amphoric breath sound (bronchial breathing)—as bronchopleural fistula is a common cause of hydropneumothorax (pleural effusion rarely may have bronchial breath sound),
- (ii) Succussion splash (absent in pleural effusion),
- (iii) Tinkling sounds (absent in pleural effusion), and
- (iv) Positive coin sound in upper chest (absent in pleural effusion).

So, describe the symptomatology and signs with special reference to above points to prove the rationality of your diagnosis.

Aetiology of hydropneumothorax :

1. Rupture of a subpleural tuberculous focus or cavity (commonest cause).
2. Rupture of subpleural lung abscess (staphylococcal).
3. Chest injury (penetrating).
4. Acute pulmonary infarction.
5. Following cardiac surgery.
6. During aspiration of fluid in pleural effusion (iatrogenic).
7. Secondary infection of pneumothorax, specially after water-seal drainage.

- * Actually No. 2 produces pyopneumothorax (pus and air), and No. 3 haemopneumothorax (blood and air).

How shifting dullness is elicited ?

Due to the presence of free fluid and air within the pleural cavity, dullness (fluid) shifts from one place to other with the change of posture of the patient (similar mechanism as ascites).

Clinical method :

1. In **sitting position** of the patient, percussion is done in the conventional method in anterior chest wall along the MCL. In hydropneumothorax, the lower part of the affected chest will be stony dull and the upper part tympanitic.
2. Now the percussion is done In **lying down position** when the fluid gravitates in the dependent part and the air comes in front. Percussion along the MCL in anterior chest wall in lying down position will elicit tympanitic resonance all over the anterior chest. So, the lower part of anterior chest which was dull on percussion in sitting position becomes tympanitic in lying down position, i.e., dullness shifts.
3. The same manoeuvre can be done in the back (always after doing the percussion in sitting position) when the patient lies in prone position.

* Alternative method—Few clinicians follow this manoeuvre. First, percussion of the back in sitting position of the patient is done and the patient is then instructed to bend forward in the same sitting position. Percussing again in the back will reveal change of fluid level (which goes up) in forward bending position of the patient.

Demonstrate succussion splash (Hippocratic succussion) :

By the method of percussion, the upper border of dullness is detected in the lateral chest wall along the MAL, in sitting position of the patient. Now the diaphragm of stethoscope is placed on the air-fluid level and the patient is shaken from side to side vigorously. A splashing sound (like splashing sound of intact coconut) is audible with every jerk. Sometimes, the sound can be heard without the help of stethoscope (unaided ear, i.e., ear placed over the chest wall and the patient is shaken from side to side). The stethoscope may also be placed in the anterior or posterior chest wall at the air-fluid level to demonstrate succussion splash. Succussion splash in the chest is always pathological and is found in :

1. Hydropneumothorax or pyopneumothorax.
2. Herniation of stomach in the thorax.
3. Rarely, over a large cavity in the lung partially filled with exudate.

How to elicit the horizontal fluid level ?

This is classically found in hydropneumothorax. In sitting position of the patient, percussion is done from above downwards in the front (along MCL), lateral chest wall (along MAL) and back (along scapular line) in the conventional method. During percussion from above downwards, a point of dullness is reached in the front, lateral chest wall and back where markings are given by the skin pencil. These three points are joined transversely and a horizontal line is drawn encircling the affected chest wall. This is the upper horizontal border of fluid level.

N.B. : Ellis's S-shaped curve in pleural effusion does not exist today. Actually, it is a radiological observation (highest level of fluid in axilla). Previously the lesson was that the highest level of dullness (elicited by percussion) is present in axilla (so called Ellis's S-shaped curve) though it is not true. The upper limit of fluid in pleural effusion is horizontal like hydropneumothorax but it is not easily demonstrable.

In hydropneumothorax, the difference in the note of percussion (from above downwards, it is tympanitic to stony dullness) is very distinct and clear cut in comparison to pleural effusion (from above downwards, it is resonant note to stony dullness). So the term 'horizontal fluid level' is classically used in hydropneumothorax.

From which ICS, the axilla starts ?

The 4th intercostal space (ICS).

Surface marking of 'root or hilum of the lung' :

It is the space in between the spine and vertebral border of scapula opposite to T₄, T₅ and T₆ vertebrae on both sides. In health, bronchovesicular breath sound is audible here.

Purpose of percussion in clinical practice :

It is done to note the resonance or dullness of an organ, and also to know the :

1. Content (state of underlying tissue i.e., whether solid organ, fluid or gas is present),
2. Contour (topographical percussion), and
3. Depth (usually by percussion, we get an idea upto a depth of 2 inches).

* Percussion indirectly diagnoses tenderness (e.g., empyema thoracis, liver abscess).

** A lesion of < 2 cm diameter does not produce change in the percussion note.

Purpose of percussion over chest :

1. Conventional or general percussion,
2. Liver or cardiac dullness,
3. Coin percussion,
4. Shifting dullness,
5. Tidal percussion, and
6. Traube's space percussion.

Three cardinal rules of percussion :

1. Percuss from resonant to dull area, or more resonant to less resonant area.
2. Pleximeter finger should be placed parallel to the border of the organ to be percussed and the line of percussion should be perpendicular to that arbitrary border.
3. Heavy percussion for deeply placed viscera (e.g., upper border of liver) and light percussion for superficial viscera (e.g., lower border of liver).

* The art of respiratory medicine remains in performing 'percussion'. The method of percussion was first described by Leopold Auenbrugger (1722-1809) when he was a medical student.

How far you can go during conventional percussion in the chest ?

In the MCL, MAL and scapular line, the lower border of lung extends upto 6th rib, 8th rib and 10th rib respectively. The lower limit of pleura is present two-spaces downwards than the lung, i.e., at 8th rib, 10th rib and 12th rib respectively. These are true in a patient with quiet respiration.

So, conventionally one should percuss :

1. Along MCL - upto 7th ICS (the space just above the lower limit of pleura).
2. Along MAL - upto 8th ICS (upper border of spleen lies at 9th rib).
3. Along scapular line - upto 11th ICS (the space just above the lower limit of pleura).

Basic principles for conventional percussion over the chest :

1. The dorsal aspect of middle phalanx of the middle finger of left hand (the PLEXIMETER FINGER) is kept tightly over the chest wall (i.e., over intercostal spaces), and is struck suddenly with the tip of the middle finger of right hand (the PERCUSSING or PLEXOR FINGER) perpendicularly (with 'hammer effect')—mediate percussion. The entire movement is delivered from the wrist joint, and not from the elbow. Strike once or twice on the pleximeter finger (avoid repeated heavy striking). The other fingers of the left hand should not touch the chest wall.
2. Ask the patient to sit. Direct percussion is done over bones and indirect percussion for other areas. First percuss the clavicle (direct percussion) and then the anterior chest wall along MCL. The first palpable interspace in anterior chest wall is 2nd ICS. *Keep the hands by the side in resting position while percussing the anterior chest wall along MCL. During percussion of axilla along MAL, patient's hands are kept over his head. While percussing the back, cross the patient's hands over the knees or shoulders* (this position wide apart the scapulae), and percuss in a bat's-wing or fish-bone pattern. Lastly, *percuss the apex of the lung from the back of the patient keeping the patient's hands by the side* (read Kronig's isthmus from page 84).
3. Always percuss from above downwards and compare the note on the identical site on the opposite side of chest. All the areas should preferably be percussed in sitting position of the patient.

* While percussing the left side of anterior chest wall, always follow the left MCL (do not deviate laterally, because the most outer border of cardiac dullness lies 1/2 inch inside the left MCL). One may start percussion from apparently healthy side, or the right side of the chest (if it is not possible to detect the diseased side from history). Use light percussion for anterior chest and heavy percussion for posterior chest. Never percuss over ribs.

Format of percussion over the chest :

- (A) Anterior chest wall :
1. Over the clavicle (direct percussion)
 2. Infraclavicular area (clavicle to 3rd rib)
 3. Mammary area (3rd to 6th rib)
 4. Inframammary area (6th rib to 7th ICS).

Both the sides of the chest should be percussed with special reference to—

Right side

1. Conventional percussion
2. Shifting dullness
3. Liver dullness
4. Coin percussion
5. -

Left side

1. Conventional percussion
2. Shifting dullness
3. Cardiac dullness
4. Coin percussion
5. Traube's space percussion

- (B) Lateral chest wall (right and left side)
 1. Axillary area (4th ICS to 6th rib)
 2. Infra-axillary area (6th rib to 8th ICS or lower costal margin)
- (C) Posterior chest wall (right and left side) :
 1. Suprascapular (above the upper border of scapula)
 2. Interscapular (between the medial border of scapula and spine-> T_2 to T_7 vertebrae->in a ■fish-bone pattern' percussion)
 3. Infrascapular (below the inferior angle of scapula-> T_7 to T_n vertebrae).
- (D) Apical (or supraclavicular area) : always done from the back (read “Kronig’s isthmus”)—right and left side.

How to count the ribs and ICS in clinical practice ?

First place the right index finger in the suprasternal notch and then go downwards till the **sternal angle** (angle of Louis) is felt as a transverse bony ridge (junction of the body of the sternum and manubrium sterni). If the finger is moved sideways, it will touch the 2nd ribs. Now count the ribs with ICS from above downwards.

Posteriorly, the ribs and ICS are counted from below upwards (difficulty faced in a muscular body or in an obese patient). If the arms rest by the side of the body, the inferior angle of scapula lies at the level of T_7 vertebra (or the 7th rib) — this may be of some help in counting ribs or ICS in the back.

* **Angle of Louis** or sternal angle (i.e., the level of 2nd costal cartilage) corresponds with the disc space between T_4 and T_5 vertebrae posteriorly, and anteriorly it is the landmark for tracheal bifurcation.

Traube's semilunar space and its importance :

This is a triangular space or semilunar topographic area in the left lower chest anteriorly which is bounded by,

Laterally - Left midaxillary line

Above - Left dome of diaphragm and left lung i.e., the 6th rib.

Below - Left lower costal margin.

Detection : The triangle is percussed during normal breathing across one or more level from its medial to lateral margin, i.e., from xiphisternum to left midaxillary line across 6th and 7th intercostal space (Barkun's method).

Importance : Normally the space is tympanitic on percussion as **it contains the fundus of the stomach**. The tympanicity of Traube's space (after Ludwig Traube) is lost in :

- a) Carcinoma involving the fundus of the stomach or full stomach.
- b) Left-sided pleural effusion.
- c) Enlarged left lobe of liver.
- d) Massively enlarged spleen or ruptured spleen.
- e) Situs inversus totalis (Traube's space is present on the right side).
- f) Achalasia cardia (often the fundal gas is absent).

* Traube's space is shifted upwards in fibrosis or collapse of the left lower lobe of lung and left-sided diaphragmatic palsy.

** Traube's space tympanicity is retained in pericardial effusion.

Shifting of upper border of liver dullness :

To delineate the upper border of liver dullness, percuss the anterior chest wall along right MCL from above downwards. Normally the **upper border of liver dullness is present in right 5th ICS at MCL**.

- (A) Lowered or obliterated :
 1. Emphysema.
 2. Pneumothorax (right-sided).
 3. Perforation of abdominal hollow viscus e.g., perforation of peptic ulcer.
 4. Chilaiditi's syndrome (interposition of colon between liver and diaphragm—an anatomical variation).
 5. Cirrhosis of liver (liver becomes small).
 6. Viscerptosis of liver.

(B) Elevated :

1. Amoebic or pyogenic liver abscess.
2. Subdiaphragmatic abscess (right).
3. Diaphragmatic palsy (right).
4. Pleural effusion (right).
5. Basal pneumonia (right).
6. Increased intraabdominal tension due to ascites or pregnancy.

* The upper border of liver dullness is present in right 7th and 9th ICS when percussed along midaxillary and scapular line respectively.

Where do you beat in clavicular percussion ?

It is a **direct percussion** (clavicle acts as pleximeter finger) done by right middle finger,

- (i) Over the most prominent part of clavicle, or
- (ii) Over the medial 1/3 rd of the clavicle, just lateral to its expanded medial end.

During percussion, stretch the overlying skin downwards with the left thumb so that the percussing finger does not slip over the clavicle. It is a light percussion. Few clinicians advise to place pleximeter finger over clavicle to reduce pain while percussing.

What is tidal percussion ?

While percussing during delineating the lower border of lung, there is difference in resonance by one space in full inspiration. It happens to be due to the downward movement of diaphragm. Today, this method has little practical value and is only of historical interest.

Method :

The patient will sit with hands crossed over the knees or shoulders. First **percuss the lower** back along the scapular line at the end of forced expiration and then percuss the same place at the height of full inspiration. Note the increase in resonance by one space after full inspiration.

Clinical importance :

1. Paradoxical resonance i.e., there is dull note on inspiration and resonant note on expiration due to paradoxical movement of diaphragm in diaphragmatic palsy.
2. Dullness which does not change on inspiration or expiration indicates basal pneumonia, basal pleurisy or subdiaphragmatic abscess.

What is Kronig's isthmus ?

It is a small area (a band of resonance of 5-6 cm width, connecting the lung resonance on the anterior and posterior chest on each side) in the apex of the lung (supraclavicular area) which is bounded,

Medially—By neck muscles, and laterally—By the ipsilateral shoulder joint.

Anteriorly—By the clavicle, and posteriorly—By the trapezius muscle.

Kronig's isthmus is elicited by percussion over the apex of the lung (percussion done from the back of the patient), where the note is normally resonant. The area becomes dull on percussion in the presence of apical tuberculosis, Pancoast's tumour or apical pneumonia. *The pleximeter finger should be placed over the supraclavicular fossa, across the anterior border of trapezius, perpendicular to the clavicle.*

What is percussion myokymia ?

In some chronic wasting diseases e.g., pulmonary tuberculosis, if a light tap is applied over the chest (on the sternum or near the sternum), it may produce transient twitching of pectoral muscles as a result of undue irritability. This phenomenon is also known as 'myotatic irritability'.

Diagnosis of diaphragmatic palsy :

Unilateral paralysis is more common than bilateral paralysis. Unilateral paralysis commonly results from tumour (bronchogenic carcinoma), birth injury, surgery or trauma to the mediastinum. Bilateral paralysis may occur due to,

- a) High cervical cord injury.
- b) Motor neurone disease.
- c) Poliomyelitis.

- d) Demyelinating disease.
- e) Polyneuropathies (e.g., G.B. syndrome).
- f) Bilateral involvement by mediastinal lesions.

Diaphragmatic palsy is diagnosed by :

1. Dyspnoea in supine posture.
2. Abdominal movement is less in comparison to thoracic movement.
3. As a whole the respiratory excursions in chest are diminished.
4. Tachypnoea may be present with intercostal suction.
5. **Paradoxical respiration** — Depression of abdomen with inspiration, and elevation with expiration. It is best observed in unilateral palsy and is known as 'see-saw movement' of abdomen.
6. Tidal percussion (see above).
7. Counting sign — Patient can not count aloud upto 10 in one breath (sometimes, it is seen that COPD patients can not count upto 20 in one breath; often it is observed that patient of myasthenia gravis starts counting normally but the voice becomes unintelligible after crossing 10).
8. Litten's sign — Absence of indrawing of subcostal margin during inspiration.
9. Elevated hemidiaphragm in chest X-ray which is confirmed by fluoroscopy.
10. **Sniff test** (diagnostic on fluoroscopy) — When the patient is asked to sniff, there is upward motion of the affected hemidiaphragm (the unaffected diaphragm descends). Sniff test can also be confirmed by USG.

* Patient with diaphragmatic palsy will have difficulty in sniffing—a rough test for integrity of diaphragm. Bilateral diaphragmatic palsy is a cause of sleep apnoea. Treatment is done by diaphragmatic pacing or nighttime assisted ventilation.

Put your hand over lower lobe of left lung :

Place your hand in the lower part of back of the chest on left side. Actually when seen **from behind**, mainly the lower lobes are seen while a small area near the apex is occupied by the upper lobes. When seen **from front**, the upper and middle lobes on the right, and the upper lobe on the left side occupy the most. When seen **from sides** (axillary region), parts of all the lobes are accessible.

Absent shifting dullness in hydropneumothorax : is it possible ?

Shifting dullness is absent in loculated or encysted variety of hydropneumothorax. A case with small air and large fluid, or large air and small fluid may not have shifting dullness.

Diagnosis of pyopneumothorax developed from hydropneumothorax :

1. Patient will be toxic and prostrated; loss of weight.
2. Hectic rise of temperature with rigors and sweating.
3. Tachycardia as well as tachypnoea.
4. Intercostal tenderness and tenderness on percussion (patient winces with pain).
5. Skin is red, oedematous and glossy over lower part of affected chest.
6. Development of clubbing, plus
7. Signs of hydropneumothorax.

D/D you will consider in your case :

1. Pyopneumothorax.
2. Pleural effusion.
3. Herniation of stomach in the thorax.
4. A large lung abscess partially filled with exudate.

Clinical differentiation between pleural effusion and hydropneumothorax :

Theoretically there may be many differences but basic bedside dissimilarities are :

Pleural effusion :

1. Percussion note above the fluid level—Skodaic resonance.
2. Shifting dullness—Absent.
3. Coin sound in upper chest—Absent.

4. Succussion splash—Absent.
5. Classical horizontal fluid level—Present but difficult to demonstrate.
6. Breath sound and vocal resonance—Commonly diminished vesicular breath sound with diminished vocal resonance is audible though bronchial (tubular) breath sound, bronchophony and whispering pictoriloquy may be heard at the upper border of moderate effusion.

Hydropneumo thorax :

1. Percussion note above the fluid level—Tympanitic.
2. Shifting dullness—Present.
3. Coin sound in upper chest—May be present.
4. Succussion splash—Present.
5. Horizontal fluid level on percussion—Easily demonstrated.
6. Diminished vesicular breath sound with diminished vocal resonance at the upper border of fluid level. There may be bronchial (amphoric) breath sound in bronchopleural fistula.

How do you like to investigate this case ?

The investigations will be done in the line of both pleural effusion and pneumothorax already described but the X-ray features will be different in hydropneumothorax.

Chest X-ray (PA view) in **erect posture** shows :

1. Horizontal fluid level on right chest.
2. Increased radiolucency above the horizontal level which is lacking in lung markings (pneumo' component) and a homogeneous opacity below the horizontal level ('hydro' component).
3. Shifting of trachea and cardiac shadow towards the left side.
4. The left lung shows wooly opacities in apex (i.e., apical tuberculosis).

It is worthwhile to remember that in erect posture, the fluid remains below and the air above. The collapsed lung is usually hidden within the fluid and is not visible in X-ray (rarely, collapsed lung is visible paravertebrally).

Blood (R/E). sputum examination and Mantoux test are done routinely.

How will you manage a patient of hydropneumothorax ?

1. Intercostal tube drainage through water-seal drainage system is applied as done in pneumothorax. The catheter is introduced in the **dependent part** in such a way that fluid can come out through the tube (special tube with multiple perforations like 'Romodrain') and the air is absorbed or expelled in due course of time.
2. Antituberculosis chemotherapy,
3. Proper antibiotics (in case of pyopneumothorax).
4. Surgery in resistant cases with pyo- or haemopneumothorax.

Management of empyema thoracis :

Diagnosis of empyema is made by clinical features, radiology (similar to pleural effusion) and aspiration of pus by a wide-bore needle through an intercostal space over the area of maximum dullness. Then bacteriological examination of pus is done accordingly.

(A) Treatment of non-tuberculous empyema :

- a) Acute —
 - (i) Intercostal tube drainage through water-seal drainage system.
 - (ii) Proper antibiotic after sensitivity test (arbitrary use may produce resistance).
 - (iii) If pus is very thick, rib resection and clearing of empyema cavity is done; a wide bore tube helps in prolonged drainage.
- b) Chronic — it is the domain of thoracic surgeon.
 - (i) Resection of empyema sac in toto (if the underlying lung is healthy).
 - (ii) Thoracotomy and decortication of pleura (in patients with thickened pleura). Decortication means stripping of the whole of the thickened visceral pleura.

(B) Treatment of tuberculous empyema :

- a) Antituberculosis chemotherapy.
- b) Repeated drainage of pus through a wide-bore needle or tube-drainage.
- c) Rarely, surgery is necessary.

Case 11

CONSOLIDATION OF THE LUNG

What is your diagnosis ?

This is a case of consolidation of the lower lobe of left lung.

Why do you say so ?

This patient is suffering from consolidation because there is presence of :

(A) SYMPTOMS (from the history) —

- a) High fever with chill and rigor, malaise and headache for 5 days.
- b) Pain on the left side of chest, and dyspnoea for 3 days.
- c) Harassing dry cough for 2 days.

(B) SIGNS —

1. General survey :

- a) Patient looks toxic and ill, but conscious and co-operative at present.
- b) Decubitus — Propped-up at 45°.
- c) Cyanosis—Absent at present.
- d) Respiration—42/min, regular and predominantly abdomino-thoracic in type.
- e) Pulse — 86/min, regular (describe other points in pulse).
- f) Pulse : Respiration—2 : 1
- g) Herpes labialis—Present on left side.
- h) Temperature (oral)—100°F at present.
- i) Neck stiffness—Absent (to diagnose meningism).

2. Inspection :

- a) Upper respiratory tract reveals no abnormality.
- b) Absence of intercostal suction but accessory muscles of respiration are working.
- c) Absence of fullness or flattening of chest wall anywhere.
- d) Apical impulse—Seen below the left nipple, approximately 3" lateral to midline.
- e) Diminished movement on left side of chest, mainly the lower part of back and lateral aspect of chest.

3. Palpation :

- a) Restricted movement on left side of chest.
- b) TRACHEA AND APEX BEAT ARE NORMAL IN POSITION (I.E., NO MEDIASTINAL DISPLACEMENT).
- c) Vocal fremitus—Increased on left side (lower part of lateral aspect and back mainly).
- d) Friction fremitus—Present in lower part of back, on the left chest.

4. Percussion :

Woody dullness on left side of chest (lower part), mainly at the back and lateral aspect. The outline of woody dullness is irregular and limited.

5. Auscultation :

All the positive findings are present mainly on the left base (left infrascapular region).

- a) Tubular breath sound (high-pitched bronchial).
- b) Increased vocal resonance—Bronchophony and whispering pectoriloquy.
- c) Aegophony.
- d) Pleural rub
- e) Absence of rhonchi and crepitations (as lung is solidified and liquefaction has not started yet).

* The right lung seems to be normal clinically. Characteristic rusty sputum may not be found.

** The surface marking of the dullness in consolidation and collapse follows the distribution of affected lobe which is not seen in pleural effusion.

*** The involvement of lung in acute bronchopneumonia is usually patchy, bilateral and more widespread in lower lobes.

**** 'Consolidation' i.e., pneumonia is exudative solidification of lung parenchyma (alveoli).

Why do you say consolidation of the lower lobe ?

The signs are predominantly present in the back and at basal region.

Pathological 'stages of pneumonia' with auscultatory findings :

- (A) Stage of hyperaemia or congestion : Diminished vesicular breath sound with fine crepitations (indurated crepitations) — 24-48 hours.
- (B) Stage of 'Red Hepatisation' (2-4 days) : \
- (C) Stage of Grey Hepatisation (4-8 days) : J T T , , , , , f Classical features of consolidation described above
- (D) Stage of resolution : Breath sound is vesicular or rarely bronchovesicular in type. Coarse crepitations are heard (redurated crepitations) — 2-3 weeks.

* Respiratory symptoms and signs in consolidation are often absent in elderly, alcoholic, immunocompromised or neutropenic patients.

Classification of pneumonia :

Pneumonia (synonym : pneumonitis) is defined as acute inflammation of the lung parenchyma.

- (A) Main classification : *

 - 1. 'Primary' pneumonia (*Streptococcus pneumoniae* [most common], *Staphylococcus aureus*, *Haemophilus influenzae*, *Klebsiella pneumoniae* etc.)—Usually community-acquired (in normal healthy Individuals).
 - 2. 'Secondary' pneumonia (occurs when the host is weakened) —
 - (i) Acute bronchopneumonia, acute lobular or hypostatic pneumonia.
 - (ii) Benign aspiration pneumonia.
 - (iii) Suppurative or necrotising pneumonia (lung abscess)—Commonly by *Staphylococcus aureus* and *Klebsiella pneumoniae*.
 - (iv) Hospital-acquired pneumonia (nosocomial).
 - (v) In immunocompromised host (*Pneumocystis jiroveci* infection in AIDS).

- * Lung abscess may be a part of 'primary' pneumonia.
- (B) Empiricist's classification (most commonly used) :
 - 1. Community-acquired pneumonia (*Streptococcus pneumoniae*, *Haemophilus influenzae*).
 - 2. Hospital-acquired pneumonia (nosocomial)—Mainly by aerobic gram-ve bacilli, anaerobes.
 - 3. Pneumonia in immunocompromised host—*Pseudomonas aeruginosa*, *Pneumocystis jiroveci*.
- (C) Anatomical classification :
 - 1. Lobar.
 - 2. Lobular (bronchopneumonia, if bilateral).
 - 3. Segmental (benign aspiration pneumonia).
- (D) Aetiological classification :
 - 1. Infective—Bacterial, viral, rickettsial, yeast and fungi, protozoa (*Pneumocystis jiroveci*).
 - 2. Chemical agents—Irritant gases, aspiration of vomitus, kerosine oil etc. (kerosine oil produces 'lipoid pneumonia').
 - 3. Physical agents—Irradiation.
 - 4) Hypersensitivity reaction—SLE, Stevens-Johnson syndrome, Loeffler's syndrome.
- (E) Radiological classification :
 - 1. Alveolar or air space pneumonia (pneumococcal mainly)— air bronchogram in chest X-ray.
 - 2. Bronchopneumonia (*Haemophilus influenzae* etc.)— atelectasis is seen in chest X-ray.
 - 3. Interstitial (virus, mycoplasma etc.)—Reticular or reticulonodular pattern in chest X-ray.
- (F) Behaviorist's classification : 1. Easy, and 2. Difficult : non-resolving.

* Now-a-days, ventilator-associated pneumonia (VAP) with MDR and non-MDR pathogens pose problem in treatment (MDR = multiple drug resistance).

Characteristics of viral pneumonia :

- 1 Constitutional symptoms like anorexia, headache, myalgia, malaise are predominant (commonly due to influenza, parainfluenza and respiratory syncytial virus). Fever and toxemia usually precede respiratory symptoms.
- 2 Cough may be absent initially and sputum is mucoid in nature.
- 3 Haemoptysis, pleuritic chest pain and pleural effusion are rare.
- 4 **Spleen may be palpable** in the first week; tachypnoea may be absent.
- 5 **Paucity of physical signs** in chest but may develop later and seldom gross.
- 6 WBC count is usually normal.

7. X-ray—Usually do not correspond to lobes or segments, which is typical of consolidation. Reticular or reticulonodular patterns may be seen.
8. Failure of rapid resolution or response with antibiotics.

Absence of physical signs of consolidation in lobar pneumonia : is it possible ?

Yes. The possibilities are :

1. Deep seated consolidation.
2. Associated pleural effusion or empyema thoracis.
3. When the bronchus is not patent (exudate fills the bronchus along with the alveoli) — As seen in 'massive pneumonia' caused by *Klebsiella pneumoniae* (Friedlander's pneumonia).

* Sometimes in hypostatic pneumonia, the classical signs of consolidation are lacking.

What is massive consolidation ?

It is the consolidation involving more than one lobe.

Appearance of physical signs of consolidation :

Usually appear within 2 days.

Usually disappear within 2 weeks.

Appearance of radiological signs of consolidation :

Opacity in X-ray appears within 12-18 hours of the onset of illness.

Radiological opacity usually disappears within 4 weeks.

How to suspect development of effusion with pneumonia ?

Development of effusion with pneumonia is known as 'synpneumonic' pleural effusion. When the effusion appears after resolution of pneumonia, it is called 'postpneumonic' pleural effusion. Together, they are known as 'parapneumonic effusion'. These terms are going to be obsolete now-a-days.

Persistence of fever, stony dullness on percussion and shifting of mediastinum make the clinician suspicious about the development of pleural effusion. Radiology (obliteration of costophrenic angle in chest X-ray) further supports the diagnosis.

Causes of 'delayed resolution' of pneumonia :

Delayed resolution means when the physical signs persist for > 2 weeks and the radiological features persist for > 4 weeks after proper antimicrobial treatment. The possibilities are :

1. Inappropriate chemotherapy like incomplete treatment, suboptimal dose, wrong choice of antibiotics, drug resistance, irregular intake of drugs.
2. Complicated by pleural effusion, suppurative pneumonia or empyema thoracis.
3. Decreased host resistance like diabetes mellitus, chronic alcoholism, agranulocytosis, lymphoma, leukaemia, multiple myeloma, corticosteroid therapy, hypogammaglobulinaemia, AIDS etc; elderly and debilitated patient.
4. Tuberculous pneumonia (needs specific antituberculosis chemotherapy).
5. Partial obstruction of bronchus by a foreign body like dentures in the elderly or **bronchogenic carcinoma** (common in clinical practice).
6. Fungal or atypical pneumonia.
7. Pneumonia resulting from connective tissue diseases, pulmonary infarction etc.
8. Recurrent aspiration in achalasia cardia.

* Common causes of 'non-resolution' of pneumonia are inappropriate chemotherapy, bronchogenic carcinoma, diabetes mellitus or pneumonia caused by virulent organism (e.g., staphylococcal pneumonia).

Possible causes of recurrent pneumonia :

Chronic bronchitis, SLE, multiple myeloma, diabetes, AIDS, recurrent aspiration in achalasia cardia or pharyngeal pouch or scleroderma, bronchiectasis, bronchogenic carcinoma, cystic fibrosis, sequestration of the lung or foreign body aspiration.

Is it possible to have shifting of trachea to the same side in pneumonia ?

In pneumonia, trachea remains in the midline. Rarely, there may be same-sided shifting of trachea in massive pneumonia producing 'collapse consolidation' (may be from bronchogenic carcinoma).

Predisposing factors to pneumonia :

1. Cigarette smoking (strongest independent risk factor).

2. Upper respiratory tract viral infection, followed by streptococcus pneumoniae infection.
3. Excess of alcohol.
4. Immunosuppression.
5. I.V drug abusers.
6. Bronchial obstruction (malignancy), bronchiectasis.
7. Old age.

When will you advise hospitalisation in a patient of pneumonia ?

Hospitalisation is advised in :

1. Elderly patients (over 60 years of age) and with,
2. Severe dyspnoea with hypoxia (tachypnoea with respiratory rate > 30/min).
3. Central cyanosis.
4. Fall of diastolic BP (< 60 mm of Hg.).
5. Empyema, meningism, meningitis.
6. Altered mental status, delirium.
7. Shock.
8. Underlying serious cardio-pulmonary diseases, diabetes mellitus, uraemia.
9. Hypoxia (PaO_2 < 60 mm of Hg), leucopenia (< 4000/mm³) or leucocytosis (> 20000/mm³ e.g., in empyema), high blood urea or culture +ve pneumonia.

* The patient is referred to ITU (Intensive Therapeutic Unit) in No. 6, 7, 8, 9 and severe acidosis.

What are the accessory muscles of respiration ?

Normal muscles of respiration are intercostals (external and internal) and diaphragm. Accessory muscles are not required for respiration in health; they are required to assist breathing in respiratory embarrassment, and may therefore be active and prominent. They are :

- | | |
|-----------------------------|-----------------------|
| 1. Alae nasi (nasal flare). | 5. Trapezius. |
| 2. Sternomastoid. | 6. Pectoralis major. |
| 3. Scalenii. | 7. Serratus anterior. |
| 4. Latissimus dorsi. | 8. Abdominal muscles. |

Common causes of 'intercostal suction' :

Normally intercostal spaces expands and draw-out in inspiration. It is the drawing-in of the intercostal spaces with inspiration and is commonly seen in,

- | | |
|--|---|
| 1. Foreign body within larynx or trachea. | 4. COPD. |
| 2. Diphtheria. | 5. Bronchiolitis (in infants and children). |
| 3. Oedema of the glottis (in anaphylaxis). | 6. Bilateral diaphragmatic palsy. |

* Produced as a result of positive atmospheric pressure pressing the lower intercostal spaces in the face of negative intrathoracic pressure due to failure of air entry in respiratory tract.

What are the D/D you will consider in your case ?

- (A) Pleural effusion ;
- a) Gradual onset.
 - b) Dry cough may be present; patient does not look toxic.
 - c) **Mediastinal shifting to opposite side, stony dullness on percussion.**
 - d) Diminished vesicular breath sound or absent breath sound with diminished or absent vocal resonance. Usually there is no added sound.
- (B) Collapse of the lung :
- a) Cause is detected from the history e.g., aspiration, trauma, post-operative events etc.
 - b) Patient does not look toxic.
 - c) **Mediastinal shift is towards the affected side** with flattening of chest wall and aggregation of ribs.
 - d) Impaired resonance on percussion.
- (C) Bronchogenic carcinoma :
- a) Age is higher; male, smoker commonly.
 - b) Clubbing.
 - c) Cervical lymphadenopathy.
 - d) Emaciation.
 - e) **Usually no shifting of mediastinum unless complicated by pleural effusion.**
 - f) Localised crepitations over the mass.

- (D) Pulmonary tuberculosis :
- Gradual onset of weight loss, anorexia, evening rise of temperature, night sweats.
 - Physical signs are mainly present in apical region, if there are any.
 - Absence of high rise of temperature or rigor.
 - Classical signs of consolidation are absent.
- (E) Pulmonary infarction (thromboembolism) :
- Sudden onset of chest pain with dyspnoea.
 - Pyrexia is usually less.
 - There may be evidence of thrombophlebitis in legs.
 - Cough is not troublesome.
 - Patient is not toxic.
 - Haemoptysis is much more common.

Causes of haemoptysis :

- | | |
|---|---|
| <p>(A) Pulmonary :</p> <ol style="list-style-type: none"> Bronchitis (acute and chronic). Pulmonary tuberculosis. Bronchiectasis. Lung abscess. Bronchogenic carcinoma. Pneumonia. Pulmonary thromboembolism. Lupus pneumonitis (i.e., from SLE) Foreign body in bronchus or lung. <p>(B) Cardiovascular :</p> <ol style="list-style-type: none"> Mitral stenosis. Acute left ventricular failure. Primary pulmonary hypertension. Eisenmenger's syndrome. | <p>(C) Coagulation disorder :</p> <ol style="list-style-type: none"> Haemorrhagic diathesis (leukaemia, haemophilia). Anticoagulant therapy. <p>(D) Miscellaneous :</p> <ol style="list-style-type: none"> Vasculitis (Wegener's granulomatosis). Arteriovenous malformation. Trauma. Aspergilloma ('fungus ball'). Goodpasture's syndrome. After bronchoscopy/lung biopsy. Systemic hypertension. Pulmonary haemosiderosis. Hydatid disease of lung. Idiopathic. |
|---|---|

* No 1, 2, 3, 5, 6, 10 are common causes of haemoptysis. Worldwide, chronic bronchitis is possibly the commonest cause of haemoptysis. Recurrent haemoptysis is commonly due to chronic bronchitis, bronchiectasis, bronchogenic carcinoma and pulmonary thromboembolism. Sources other than lower respiratory tract like nasopharyngeal bleeding or laryngeal carcinoma may be a cause of haemoptysis.

What is massive haemoptysis ?

Variably defined as coughing out of blood of > 100-600 ml/24 hours is 'massive' one and potentially lethal due to combined effect of blood loss as well as asphyxia.

Table 5 : Bedside differentiation between haemoptysis and haematemesis

Features	Haemoptysis	Haematemesis
1. Definition	Coughing out of blood	Vomiting out of blood
2. Symptoms	Symptoms of pulmonary and CVS diseases	Symptoms of upper G.I. tract diseases
3. Content and colour	Blood is mixed with sputum; bright red in colour and 'frothy' from admixture of air	Blood is mixed with food particles, may be coffee-ground in colour
4. Premonitory Symptoms	Premonitory symptoms like cough, salty sensation or tickling sensation in throat	Nausea, vomiting, retching, fainting, abdominal discomfort, perspiration
5. Melaena	Melaena does not occur (very rarely may occur, if blood is ingested)	Usually followed by melaena on the next day
6. Amount	Relatively less; shock is rare	Huge in amount; patient may be in impending shock
7. Reaction	Alkaline (blue litmus remains unchanged)	Acidic (blue litmus turns red)

Special signs in a 'cavity' within the lung :

Pulmonary cavity is an area of liquefaction necrosis within the lung (may remain empty or filled with secretion) and is usually communicating with a patent bronchus.

1. Post-tussive crepitations (crepitations evoked by coughing).
2. Post-tussive suction (high-pitched sucking sound audible during the phases of long inspiration following a bout of cough)—Hallmark sign of a thin-walled superficial, compressible cavity connected with the patent bronchus.
3. Cracked-pot sound by percussion over (a tympanitic note) empty communicating cavity (mimics the sound produced by percussion over a cracked pot).

Findings over a superficial, big, empty cavity connected with a patent bronchus :

1. Slight reduction in chest movement on affected side.
2. Absence of mediastinal displacement.
3. Increased vocal fremitus.
4. Percussion—Hyperresonant or tympanitic note when filled with air (empty); impaired note when filled with exudate.
5. Cavernous breath sound (low-pitched bronchial) with bronchophony and whispering pectoriloquy.
6. Coarse crepitations (specially when filled with exudate); may be post-tussive.

* Physical signs are absent in deep-seated cavity.

Common causes of pulmonary cavity :

1. Pulmonary tuberculosis (commonest), anaerobic infection in lung (lung abscess).
2. Lung abscess.
3. Necrotic degeneration of bronchogenic carcinoma (i.e., cavitating lung cancer).
4. Bronchiectasis.
5. Pseudocavity in large emphysematous bulla or lung cyst.
6. Wegener's granulomatosis, rheumatoid arthritis, pulmonary infarction, fungal cavity.

Vocal resonance (voice sounds) and its classification :**(A) DEFINITION:**

This is the auscultation of laryngeal vibrations transmitted to the chest wall and nothing but the auscultatory homologue of vocal fremitus.

(B) METHOD OF ELICITATION :

1. The patient will repeat 'one, one' or 'ninety-nine, ninety-nine'.
2. The patient will speak in a constant tone and voice (the depth and intensity of voice remaining the same); the voice should be clear.
3. Both sides of the chest are auscultated area by area, **comparing with the corresponding site on the other side**. Auscultation starts from above downwards in the front, sides and back of the chest.
4. It is better to start from the apparently healthy side. Do not auscultate over clavicle, sternum and scapula.

(C) Classification :**I) Quantitative change —**

1. Diminished — Pleural effusion, pneumothorax, hydropneumothorax, thickened pleura, fibrosis of the lung, collapse with obstructed bronchus, emphysema etc.

2. Increased —

(1) **Bronchophony** — Seems to appear from the **earpiece of stethoscope**.

(ii) **Whispering pectoriloquy** —

a) Patient whispers.

b) Increased sound is heard clearly or distinctly, i.e., syllable by syllable (pectoriloquy).

c) It seems to be spoken right into the **auscultator's ear**.

Bronchophony and whispering pectoriloquy are classically heard over consolidation. It is

also heard in collapse with patent bronchus, over a superficial empty cavity, sometimes above the level of pleural effusion, and in an open pneumothorax.

II) Qualitative change —

Aegophony — This is a qualitative change in vocal resonance with nasal intonation mimicking the bleating of a goat. It is classically found over consolidation and sometimes, above the level of pleural effusion. Though the speech remains intelligible, the removal of low-pitched elements of the sound explains the nasal bleating quality of aegophony (i.e., there is selective transmission of high-pitched components of the sound).

* The mechanism of production of quantitative increase in vocal resonance and that of bronchial breath sound is same, and naturally both are present in a single patient.

What is normal vocal resonance :

1. The sound seems to be produced at the **chest piece of stethoscope**,
2. Heard as indistinct rumble, and
3. Individual syllables are indistinguishable.

Complications of pneumonia :

(A) Pulmonary :

- | | |
|--------------------------------|----------------------------------|
| 1. Delayed resolution. | 5. Pyopneumothorax. |
| 2. Lung abscess or cavitation. | 6. Pneumatocele.* |
| 3. Pleural effusion. | 7. Fibrosis of the lung, rarely. |
| 4. Empyema thoracis. | 8. Respiratory failure. |

(B) Neurological :

- | | |
|----------------------|----------------------|
| 1. Mental confusion. | 3. Meningitis. |
| 2. Meningism. | 4. Cerebral abscess. |

(C) CVS :

- | | |
|------------------|------------------------------------|
| 1. Myocarditis. | 3. Pericarditis. |
| 2. Endocarditis. | 4. Peripheral circulatory failure. |

(D) G. I. tract : Meteorism (distended tense abdomen), hepatitis.

(E) Musculo-skeletal : Septic arthritis.

(F) Septicaemia, herpes labialis, thrombophlebitis, uraemia, ARDS, multi-organ failure.

* Multiple thin-walled cystlike lesions in staphylococcal pneumonia.

What is atypical pneumonia ?

There is a group of pneumonia which do not behave like lobar pneumococcal pneumonia (typical pneumonia) and do not respond to penicillin. This type of 'atypical' pneumonia is not severe and is caused by Chlamydia psittaci (psittacosis), Coxiella burnetii (Q fever), Legionella pneumophila and Mycoplasma pneumoniae. Predominant features are dry cough, constitutional symptoms, abnormality in chest X-ray in spite of lack of physical signs in chest.

How will you investigate pneumonia ?

1. Blood examination — TC, DC and ESR. TC shows polymorphonuclear leucocytosis. Persistence of leucocytosis signifies lung abscess or empyema formation, and absence of leucocytosis carries a bad prognosis (atypical pneumonia). Toxic granulation within the neutrophils may be present in bacterial pneumonia. ESR tends to be high. Blood culture may be helpful.
2. Sputum examination :
 - (i) For identification of organism, and
 - (ii) Detection of sensitivity to antibiotics.

— Always an attempt should be made to establish a microbiological diagnosis, if possible. Administration of nebulised hypertonic saline often helps in expectoration. Many a time false positive results come out (non-pathogenic organism from oropharynx). In our country, sputum should always be examined for identification of AFB. So, smear examination of sputum by both Gram and Ziehl-Neelsen stains, and culture and sensitivity test of sputum should be done.

3. X-ray chest (PA and lateral view)—This is helpful for confirmation of diagnosis. Homogeneous opacity is seen involving segments, part of a lobe or lobes of lung. Rarely, reticular patterns may be seen (viral, *Mycoplasma pneumoniae*). Pleural effusion or empyema may be present. Follow-up X-ray after 7-10 days is important to assess the response to therapy and for diagnosis of development of any complication.

Sites of lung involvement in pneumonia are as follows—Right lung is involved more because of its continuity with the trachea; apical area is affected more in tuberculosis or *Klebsiella pneumoniae* infection; basal segments of lower lobes in sitting-up position, and posterior segment of upper lobe and superior segment of lower lobe are mostly affected in a recumbent patient.

4. Blood culture—Sometimes this is done in severe pneumonia, specially when the sputum examination is negative. PCR tests are available for a number of pathogens.
5. Serological tests—For the diagnosis of *Mycoplasma pneumoniae*, *Coxiella burnetii*, *Legionella pneumophila* or viral infections; a four-fold rise in antibody titre repeated after 10 days suggests a recent infection. Pneumococcal antigen can be detected in pneumococcal pneumonia.
6. Nose and throat swabs—May be cultured for viruses or examined under electron microscope.
7. Pleural fluid—If there is formation of parapneumonic effusion, the fluid may be aspirated for microbiological examination.
8. Pulmonary function test (PFT) and arterial blood gas analysis (ABG)—Usually hypoxia with hypocarbia is seen (type I respiratory failure). It is done in the management of seriously ill patients in anticipation of assisted ventilation.
9. CT scan of thorax, if an obstructing tumour is suspected.
10. Bronchoalveolar lavage (BAL) or transtracheal aspiration.
11. Bronchoscopy—May be done whenever there is evidence of an obstructing tumour or foreign body.
12. Lung biopsy—Rarely needed. Sometimes, open lung biopsy is done in patients who are immunocompromised, e.g., *Pneumocystis jiroveci* infection in AIDS.

Line of management of pneumonia :

1. General—Rest in bed, good nutritious diet (high calorie with plenty of fluids).
2. Specific—Antibiotics; usually given after sputum examination. If sputum is unobtainable or unhelpful, start with erythromycin 500 mg QDS, orally. In hospital-acquired pneumonia and pneumonia in immunocompromised host, a third-generation cephalosporin should be started.
3. Symptomatic (hospitalisation, wherever indicated)—
 - a) O₂ inhalation in high concentration for cyanosis.
 - b) Cough suppression by methadone, pholcodine etc. in harassing non-productive cough; otherwise, expectoration should be encouraged.
 - c) Hydrocortisone for peripheral circulatory failure.
 - d) I.V fluid infusion for dehydration.
 - e) Warmth, counterirritants and analgesics (e.g., paracetamol) for pleural pain.
 - f) Inj. diazepam (use cautiously) for delirium.
 - g) Assisted ventilation in respiratory failure.
 - h) Postural drainage and chest physiotherapy in cases of suppurative pneumonia.

Conclusion :

Mediastinum (trachea and apex beat) is not shifted in consolidation. Fever (may have chill and rigor), cough (initially dry and painful, later on productive), haemoptysis (rusty sputum or frank haemoptysis), gross anorexia, headache and bodyache, pleuritic chest pain, tachypnoea with herpes labialis are common mode of presentations.

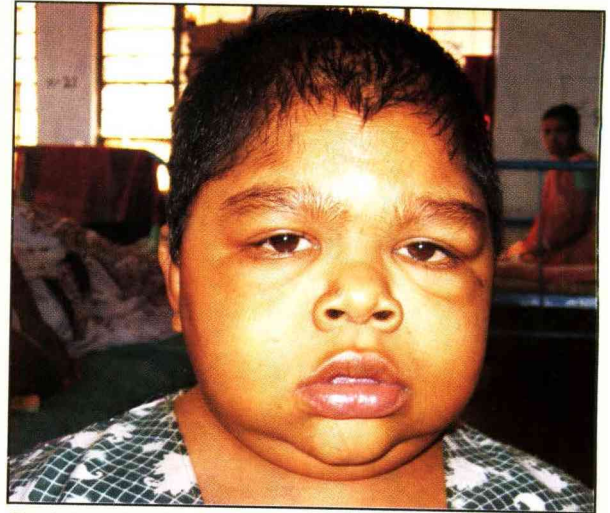
The **hallmarks of diagnosis** are :

1. Absence of mediastinal shifting.
2. (Woody) dullness on percussion on the affected side.
3. Tubular breath sound, bronchophony and whispering pectoriloquy with aegophony on the affected side.

Clinical Atlas



Thalassaemia with frontal bossing, depressed bridge of the nose, malar prominence and mild icterus



cretinism showing dwarfism, depressed bridge of the nose, broad flat nose with big nostrils, scanty hairs in scalp, thick lips with a dull, idiotic and expressionless face



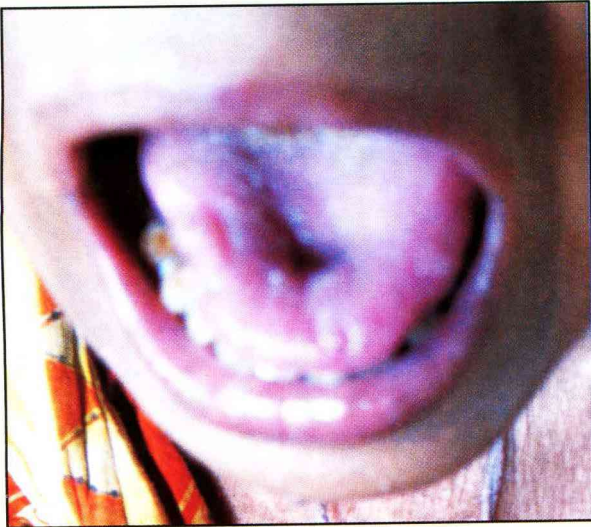
Expressionless putty face in myxoedema



Acromegaly characterized by prognathism, enlargement of face and 'spade-shaped' hands



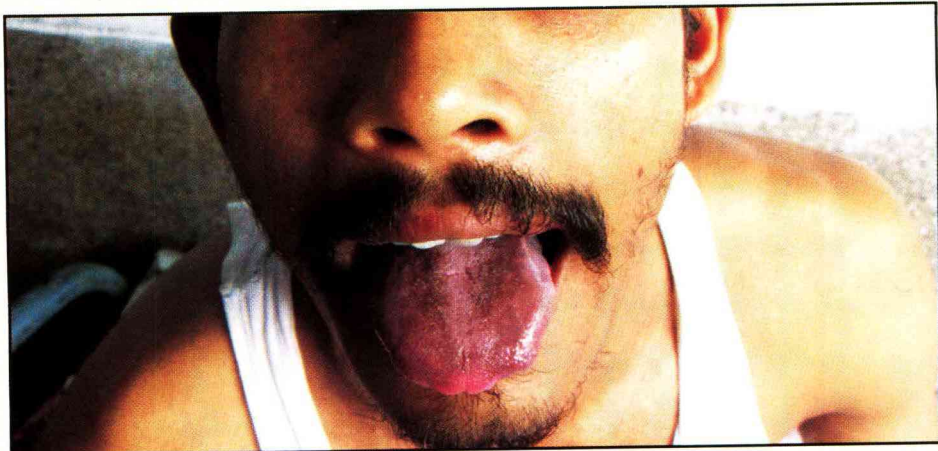
Thyrotoxicosis with exophthalmos and staring look



Central cyanosis in acute severe asthma



Suffused bulbar conjunctiva and increased redness of lower palpebral conjunctiva in **polycythemia** from cyanotic congenital heart disease



Bluish-black tongue in **methaemoglobinemia**



Gum hypertrophy in acute myeloid leukaemia



Angioneurotic oedema of face with swollen lips

Case 12

BRONCHIAL ASTHMA

What is your diagnosis ?

It is a case of bronchial asthma with recent acute paroxysmal attack.

Why do you say so ?

This patient is suffering from bronchial asthma because of the presence of :

(A) SYMPTOMS (from the history)—

1. Respiratory distress for 2 years with acute exacerbation for last 3 days.
2. Cough with mucoid expectoration for last 2 years.
[H/O present illness—describe dyspnoea, fever, cough, chest pain, H/O oedema, seasonal variation etc, if any. Past history—chronic exposure to dust and smokes. H/O repeated cough and cold in the childhood. Recurrent attacks of rhinorrhoea and angioneurotic oedema present.
Personal history—smoker (12 cigarettes/day for 14 years) and bus driver by profession.
Family history—maternal uncles suffer from same type of respiratory distress.]

* Family history regarding bronchial asthma, hay fever and eczema are important in respiratory system. Occupational history e.g., exposure to coal mines, asbestos, silica dust, or H/O exposure to cotton dust, pollen, perthenium, feather, or pets (i.e., pigeon, parrot) should always be taken into account. Drugs like fi-blockers or aspirin can induce bronchoconstriction. For bronchiectasis, ask for H/O measles, influenza or whooping cough in childhood.

(B) SIGNS—

1. General survey :

- a) Decubitus—Propped-up at 45°.
- b) Cyanosis—Absent at present.
- c) Neck veins—Within normal limit (neither engorged nor pulsatile).
- d) Polycythemia—Absent (present in patients with COPD).
- e) Respiration—30/min. regular and without purse-lip respiration.
- f) Temperature—101° F (oral) at present.
- g) Pulse—90/min, regular, without any special character (describe all other points).

2. Inspection :

- a) Accessory muscles of respiration are working.
- b) Wheeze—Present.
- c) Upper respiratory tract—Hypertrophied inferior turbinate; no nasal polyp seen.
- d) Indrawing of suprasternal and supraclavicular fossae without any intercostal suction at present.
- e) Shape of the chest—Normal.
- f) Respiration— 30/min. regular in rhythm and predominantly thoraco-abdominal in type.
- g) Tracheal tug—Absent.

* Severe bronchial asthma persisting from childhood may have 'pigeon chest'.

3. Palpation :

- a) Apex beat—Can not be localised properly (due to overinflation of lung).
- b) TRACHEA—In the midline (central).
- c) Movement—Both the sides are moving simultaneously and symmetrically, and the expansion is 2" (measured by the tape).
- d) Vocal fremitus—Symmetrically reduced all over the chest (bilaterally and uniformly).
- e) Rhonchial fremitus—Present.

4. Percussion :

- a) Resonant note all over the chest (bilaterally and uniformly).
- b) Liver dullness—Present at right 5th ICS at right MCL.

5. Auscultation :

- a) Vesicular breath sound with prolonged expiration present all over the chest.
- b) Vocal resonance—Diminished uniformly all over the chest.
- c) Adventitious sounds—
 - (i) Sibillant (high-pitched), polyphonic expiratory and inspiratory rhonchi all over the chest.
 - (ii) Coarse crepitations at the bases.
 - (iii) Absence of pleural rub.

Describe the 'breathlessness' in your patient :

1. Acute respiratory distress for last 3 days though he is suffering from mild to moderate respiratory distress from time to time for last 2 years.
2. This attack is paroxysmal, and probably precipitated by respiratory tract infection.
3. Exertional at present.
4. H/O seasonal variations (increased in winters).
5. Associated with wheeze.
6. Associated with cough and there is production of mucoid expectoration.
7. No H/O pain chest.
8. No H/O PND.
9. The disease was stationary for last 2 years (non-progressive).
10. It used to relieve by inhaled drugs and taking rest.

Gradation of respiratory dyspnoea :

The Medical Research Council Classification (dyspnoea scale) is as follows :

Grade I : Dyspnoea on strenuous exertion.

Grade II : Dyspnoea on hurrying on a level ground or walking up a small hill.

Grade III : Dyspnoea is of such a degree that the person has to stop after a mile or walk slower than most people on level ground.

Grade IV : Dyspnoea appears after walking about 100 yards.

Grade V : Dyspnoea even on undressing or at rest.

* Actually, try to assess the severity by asking to what extent the lifestyle is affected.

What do you mean by wheeze and stridor ?**(A) Wheeze :**

1. High-pitched musical sound heard from a distance (specially heard by bed-partner at night).
2. *Better heard in expiratory phase.*
3. Indicates small airways obstruction.
4. Usually associated with rhonchi in the chest.

Clinical associations :

- | | |
|---------------------------|--------------------------------------|
| a) Bronchial asthma. | e) Carcinoid syndrome. |
| b) Chronic bronchitis. | f) Endobronchial disease (neoplasm). |
| c) Tropical eosinophilia. | g) GERD with aspiration. |
| d) Cardiac asthma. | |

* Bronchial asthma is associated with reversible wheeze whereas COPD with irreversible one.

(B) Stridor :

1. Low-pitched crowing or croaking sound (loud) heard from a distance.
2. *Better heard in inspiratory phase.*
3. Indicates larger airways obstruction like larynx, trachea and major bronchus.
4. Very common in children.
5. *Laryngeal stridor is a medical emergency* and urgent respiratory support may be required.

Clinical associations :

- a) Foreign body inhalation in larynx or trachea.
- b) Laryngeal oedema (angio-oedema).
- c) Diphtheria, whooping cough.
- d) Vocal cord palsy due to *recurrent laryngeal nerve paralysis* (other features of recurrent laryngeal nerve paralysis are bovine cough and hoarseness of voice).
- e) Croup (acute laryngotracheobronchitis).
- f) Acute epiglottitis.
- g) Paratracheal and subcarinal lymph nodes pressing over main bronchi (e.g., from bronchogenic carcinoma).
- h) Laryngismus stridulus in tetany.
- i) Rapidly progressive laryngomalacia. papilloma of larynx.

Classify respiratory disorders :

(A) Obstructive ventilatory defect : Bronchial asthma, COPD, bronchiectasis, cystic fibrosis; FEV₁ is grossly 1, VC 1 or normal, and FEV₁ / VC 4-

(B) Restrictive ventilatory defect: Pleural diseases, interstitial lung disease, alveolar diseases, neuromuscular disorders (myasthenia gravis, G. B. syndrome), thoracic cage abnormalities (severe kyphoscoliosis, ankylosing spondylitis, obesity); FEV₁ is slightly ↓, VC ↓ and FEV₁/VC normal or ↓

What is 'bronchial asthma'?

It is a chronic inflammatory disorder of the airways where the tracheobronchial tree shows increased responsiveness to multiplicity of intrinsic and extrinsic stimuli, producing reversible airflow obstruction (due to muscle spasm, mucosal oedema and viscid secretion), and is characterised by paroxysm of dyspnoea, cough, tightness of chest and wheeze. Bronchial asthma is of two types :

1. Episodic asthma—Early onset, extrinsic or atopic, and
2. Persistent or chronic form—Late onset, intrinsic or non-atopic types.

What is 'acute severe asthma'?

This condition was previously known as '**status asthmaticus**'. It is a severe airway obstruction where paroxysm of acute asthmatic attacks occur without any remission in between and is not controlled by conventional bronchodilators. Remission is usually attained by the use of corticosteroid and sometimes, by respiratory assistance. This is a life-threatening attack of bronchial asthma and should be treated urgently. The clinical features are :

1. Severe dyspnoea (orthopnoea) with wheeze, tachypnoea, restlessness.
2. Active accessory muscles of respiration.
3. Unproductive cough in the presence of profuse bronchial secretions.
4. Profuse sweating.
5. Central cyanosis.
6. Tachycardia.
7. Pulsus paradoxus.
8. Diminished breath sound with minimal or absence of high-pitched polyphonic rhonchi 'SILENT CHEST' (airflow may be insufficient to produce rhonchi).
9. PEF_R (peak expiratory flow rate) is < 50% of predicted or patient's best.

How to assess the severity of bronchial asthma at the bedside?

Clinical parameters of life-threatening asthma are :

1. Exhaustion and unable to speak in one breath; inability to lie supine.
2. Confusion and diminished level of consciousness.
3. Pulse rate > 120/min.
4. Pulsus paradoxus.
5. Central cyanosis.
6. Silent chest (ominous sign).
7. Hypotension.
8. Coma.
9. Unrecordable PEF_R.
10. Arterial blood gas (ABG) analysis—
 - Normal or high PaCO₂ > 5-6 kPa (37.5 to 45 mm to Hg).
 - Severe hypoxaemia, pO_a < 8 kPa (60 mm of Hg).
 - Low pH < 7.2 or high H⁺ concentration.

* It is said that tachypnoea and apparent distress may be misleading. In life threatening asthma, there may be bradycardia and arrhythmia.

What is chronic obstructive pulmonary disease (COPD)?

It is a condition where there is chronic obstruction to airflow (major site being the small airways) due to chronic bronchitis, emphysema, and chronic bronchiolitis (small airway disease). The airflow limitation is progressive and not fully reversible. Often it is known as chronic obstructive lung disease (COLD) or chronic obstructive airway disease (COAD). The nearest D/D is chronic severe bronchial asthma.

What are the alterations in breathing?

1. DYS-PNOEA—Abnormally uncomfortable subjective awareness of breathing. Dyspnoea is a subjective symptom and related to undue consciousness for the respiratory effort ('dys' means 'difficult, painful', and 'pnoia' means 'breathing').
2. ORTHO-PNOEA—Dyspnoea in recumbent (lying down) position and is of such a degree as to make the patient sit upright for relief. The patient may sleep in a chair or uses some extra pillows at night (often quantified by number of pillows e.g., 'two-pillow orthopnoea'). There is

less efficient lung movement in lying down position, and sitting gives relief to the patient by helping in descent of liver with free diaphragmatic movement as well as diminishing the venous return in a failing heart.

3. Trepopnoea—Dyspnoea only in left or right lateral decubitus position, most often in patients with heart disease.
4. TACHYPNOEA or polypnoea—Increased rate of respiration e.g., $> 20/\text{min}$ is tachypnoea (normal respiratory rate in an adult is 14-18 breaths/min). Very high respiratory rate is observed in lobar pneumonia. Many a time, it is the only clue to shortness of breath or presence of respiratory disease.
5. HYPERVENTILATION—Increased depth of respiration (ventilation in excess of metabolic needs).
6. HYPERPNOEA—Increase in rate and depth of respiration (increased ventilation due to increased metabolic needs).
7. Platypnoea or orthodeoxia—Dyspnoea only on assuming upright position, i.e., just opposite to orthopnoea. Though rare, it happens to be due to some cardiac diseases (e.g., left atrial myxoma or left atrial ball valve thrombus), massive pulmonary thromboembolism, pulmonary arterio-venous fistula, hepato-pulmonary syndrome or in selective paralysis of intercostal muscles.
8. Bradypnoea—Abnormally low respiratory rate (e.g., $< 10/\text{min}$), commonly due to opium or morphine poisoning, increased intracranial tension, myxoedema coma, carbon dioxide narcosis, hypothermia or respiratory centre abnormality.
9. Sighing respiration—It is functional in nature, and occurs mostly in young females and children. Here, deep inspiration is followed by deep expiration with a pause in between the two. It is found in the presence of relatives or parents, and never occurs during sleep or when the patient is unmindful; if associated with 'hyperventilation syndrome', the patient may complain of giddiness, black-out, tingling of extremities, circumoral numbness, palpitation, chest wall tightness with local tenderness elicited all over the chest. Restlessness, anxiety and panic attacks are common. Features of tetany (e.g., carpopedal spasm) may be manifested in some cases. Tachypnoea and occasionally tachycardia may be present. This type of hyperventilation returns to normal during sleep, and often associated with underlying anxiety or depression.
10. Cheyne-Stokes respiration—This is a form of periodic respiration where periods of apnoea (cessation of breathing for 10 seconds or more) rhythmically alternate with periods of hyperpnoea, each phase lasting for about 30 seconds, and the whole cycle is usually completed in less than 2 minutes. The rate and depth of respiration increase to a maximum (hyperpnoea), and then decrease when breathing virtually ceases (apnoea). The hypercapnoea (as a result of apnoea) stimulates brainstem centres resulting in hyperventilation drive, which washes out CO_2 resulting in depression of respiratory centre leading to apnoea. It is commonly seen in,
 - a) Healthy infants, adults during deep sleep, in high altitude (physiological).
 - b) Severe cardiac failure (acute LVF).
 - c) Uraemia.
 - d) Narcotic poisoning (opium, barbiturates).
 - e) Increased intracranial tension.
 - f) Severe cardio-respiratory embarrassment.
11. Kussmaul's breathing—Vide infra.
12. Stertorous breathing—Breathing with rattling noise in throat (i.e., fall-back of tongue, pharyngeal secretions), often seen in deep coma or in dying patients, is known as stertorous breathing.
13. Cogwheel breathing—An interrupted type of breathing pattern, seen particularly in nervous/anxious/weeping individuals.
14. Gasping respiration—Dying patients often take few deep, irregular inspirations mimicking a 'fish out of water', is known as 'gaspings' respiration. It is the terminal respiratory pattern of lower brainstem damage.

Common causes of orthopnoea, tachypnoea, hyperpnoea and hyperventilation :

(A) Orthopnoea :

1. CCF.
2. LVF.
3. Bronchial asthma.
4. COPD.
5. Massive pleural effusion.
6. Pericardial effusion (cardiac tamponade).
7. Huge ascites.
8. Bilateral diaphragmatic palsy.

(B) Tachypnoea :

1. Nervousness, exertion, anxiety.
2. Fever.
3. Acute lobar pneumonia.
4. LVF.
5. COPD (hypoxia).
6. Shock, ARDS.
7. Acidosis.
8. Hysteria.

(C) Hyperpnoea :

1. Severe exertion e.g., 100 or 200 meter race.
2. Metabolic acidosis (Kussmaul's breathing).
3. Hysteria.

(D) Hyperventilation :

1. Hysteria.
2. Pain anywhere in the body, fever, anoxia
3. Anxiety neurosis.
4. Salicylate overdose.
5. Metabolic acidosis.

What is Kussmaul's breathing (air hunger) ?

Deep sighing, rapid breathing at a regular rate and with a hissing sound (i.e. there is hyperpnoea with increased rate and depth of respiration) is known as Kussmaul's breathing. It is commonly seen in metabolic acidosis and is also known as 'acidotic breathing'. The causes are :

1. Diabetic ketoacidosis.
2. Uraemia.
3. Cerebral tumour.
4. Sometimes in hepatic coma.

Features of upper airway obstruction (e.g., laryngeal oedema) :

1. Restlessness with hyperactive accessory muscles of respiration.
2. Dyspnoea and even orthopnoea.
3. Cough (croupy cough in diphtheria).
4. Stridor.
5. Central cyanosis.
6. Intercostal suction.

Bedside diagnosis of advanced airflow obstruction (COPD or bronchial asthma) :

1. Dyspnoea and even orthopnoea (sometimes with purse-lip breathing).
2. Hyperactive accessory muscles of respiration.
3. Reduction in the length of trachea palpable above the suprasternal notch.
4. Tracheal tug (tracheal descent during inspiration; if the intercostal distance is reduced to less than 3 fingerbreaths, it indicates hyperinflation of the lungs).
5. Inspiratory excavation of the suprasternal and supraclavicular fossae.
6. Expiratory filling of neck veins.
7. Central cyanosis.
8. Intercostal suction.
9. Relative increase in antero-posterior diameter of the chest.
10. Wheeze.

* Characteristic 'tripod' position (patient sits with knee bending and flexed head placed in between knees) and Hoover's sign (paradoxical inward movement of rib cage with each inspiration) may be seen in advanced disease.

How to assess 'respiratory depression' (e.g., opium poisoning) at the bedside ?

1. Features of hypoxia (vide page 104).
2. Presence of central cyanosis.
3. Shallow respiration (expansion of the chest will be less).
4. Decreased respiratory rate (bradypnoea).
5. Diminished air entry in lungs (diagnosed by auscultation).

* **Hypoventilation** is regular, rhythmic and shallow breathing (may be due to brainstem defects, alveolar disorder, chest wall defects or massive obesity).

Breath sound over compensatory emphysema :

Usually it is harsh vesicular. Compensatory emphysema is usually unilateral.

Bedside diagnosis of emphysema :

1. Barrel-shaped chest.
2. Purse-lip respiration.
3. Non-localisation of the apex beat.
4. Diminished chest expansion.
5. Diminished vocal fremitus.
6. Hyperresonant note on percussion bilaterally.
7. Loss of both liver and cardiac dullness.
8. Breath sound will be diminished vesicular with prolonged expiration.
9. Diminished vocal resonance.
10. Usually rhonchi and crepitations are absent. Sometimes, there is wheezy rhonchi.

11. Heart sounds — Distant (muffled).

12. Liver and spleen may be palpable due to descent of diaphragm.

* Added to these 12 points, there are features of 'advanced airflow obstruction' (question above).

** **Emphysema** is the permanent overinflation of air spaces in lung distal to terminal bronchiole as well as destruction of alveolar septae. Emphysema is of four types : centriacinar, paracinar, paraseptal, and scar or irregular.

Classification of crepitations :

Crepitations are discontinuous (< 20 millisecond in duration) bubbling or crackling (wet) sound produced by the passage of air through exudate-filled bronchi, bronchioles, alveoli, big cavity, or due to sudden snapping open of the relatively stiff alveoli at inspiration.

(A) Fine crepitations (end-inspiratory) — Soft, high-pitched and <10 millisecond in duration, and is produced due to presence of exudate in the alveoli.

1. LVF (at lung bases).
2. Fibrosis of the lung.
3. Inducible crepitations of pneumonia (i.e., early stage of consolidation).
4. Fibrosing alveolitis (velcro crepitations) - See below for its mechanism of production.

* Velcro crepitations — sound resembles as if two strips of adhered velcro tapes are being separated.

(B) Coarse crepitations (biphasic)—Louder, low-pitched and < 20 millisecond in duration, and is produced due to presence of exudate in the bronchi, bronchioles, alveoli or big cavities.

1. Acute bronchopneumonia.
2. Bronchiectasis.
3. Lung abscess or pulmonary cavity.
4. Reducible crepitations of pneumonia (i.e., resolution stage of consolidation).
5. Cystic diseases of lung.
6. Pulmonary oedema (often called 'death rattle' when occurs as a terminal event).

Recent classification of crepitations :

The new terminology for crepitations is 'crackles'. Now-a-days crepitations are described by the undermentioned **two points** :

1. To note their timing in relation to respiratory cycle, and
2. Whether they are affected by coughing or not. Crepitations heard in fibrosing alveolitis are due to sudden snapping open of the relatively stiff alveoli at the end of inspiration, and are uninfluenced by coughing. Crepitations in the presence of secretions is confirmed by the observation that they either disappear temporarily or reduce in number after coughing.

* Crepitations of LVF are often silenced by bending forward.

Types of crepitations in relation to phases of respiration :

(A) Inspiratory—

1. Early—Chronic bronchitis.
2. Mid—Pulmonary oedema.
3. Late—Fibrosing alveolitis, pulmonary oedema, lung abscess, cavity within the lung..

(B) Expiratory—

Severe airway obstruction e.g., bronchial asthma.

* Bronchiectasis gives rise to biphasic i.e., both inspiratory and expiratory crepitations.

What is post-tussive crepitations ?

These are the crepitations heard after coughing and are often diagnostic of superficial tuberculous cavity.

How do we classify rhonchi ?

Rhonchi are continuous (> 80 millisecond in duration), uninterrupted musical (dry) sound produced in the narrowed bronchi or bronchioles as a result of muscle spasm, collection of viscid secretion, mucosal oedema or narrowing by an endobronchial growth.

(A) Old classification :

1. High-pitched rhonchi which are produced in smaller bronchi or bronchioles, and of squeaky quality is known as 'SIBILANT' rhonchi (end-inspiratory or early-expiratory)—Found in bronchial asthma.
2. Low-pitched rhonchi which are produced in larger bronchi, and of snoring quality is known as 'SONOROUS' rhonchi (biphasic)—Found in endobronchial growth affecting a large bronchus.

(B) Recent classification :

1. **Monophonic rhonchi** - May be inspiratory or expiratory or both, and may change in intensity with change of posture. It is produced due to narrowing of a single bronchus by tumour or foreign body (i.e., localised obstruction), which produces a single musical note.
2. **Polyphonic rhonchi** - Particularly heard in expiration and are characteristically found in diffuse airflow obstruction e.g., bronchial asthma or chronic bronchitis. They denote dynamic compression of bronchi. This is the most common type of rhonchi where the musical sound (rhonchi) contains several notes of different pitch, and results from oscillation of many large bronchi at a time.

* Few clinicians now prefer to call rhonchi as 'wheeze'.

Common causes of rhonchi :

1. Bronchial asthma, COPD.
2. Wheezy bronchitis.
3. Cardiac asthma (LVF).
4. Tropical pulmonary eosinophilia.
5. Laryngeal oedema, foreign body or tumour obstructing bronchus (fixed monophonic).

What is chronic cor pulmonale ?

It is a condition where there is right ventricular hypertrophy with or without failure resulting from diseases affecting the structure (i.e., lung parenchyma and vasculature) and / or function of the lung. This is also known as 'pulmonary heart disease' or heart disease secondary to disease of the lung.

(A) Disorders of lung parenchyma :

COPD (commonest cause), bronchial asthma, pneumoconiosis, fibrosing alveolitis, scleroderma etc.

(B) Diseases of pulmonary vessels :

Primary pulmonary hypertension, recurrent pulmonary thromboembolism, polyarteritis nodosa etc.

(C) Disorders affecting movements of thoracic cage (i.e., affecting the function of lung) : severe kyphoscoliosis, ankylosing spondylitis, neuromuscular disorder like poliomyelitis, thoracoplasty, Pickwickian syndrome (hypoventilation with extreme obesity) etc.

What is acute cor pulmonale ?

It is synonymous with acute pulmonary thromboembolism where **right ventricle dilates with or without failure**. As a result, acute pulmonary hypertension develops from massive pulmonary thromboembolism.

Features of chronic cor pulmonale :

(A) Symptoms : Cough, expectoration, breathlessness (may not be relieved by sitting up), atypical chest pain, weakness, pedal swelling, excessive day time somnolence.

(B) General survey :

1. Orthopnoea or a stooping forward position in bed: tachypnoea; purse-lip respiration.
2. Central cyanosis.
3. Pulse - Tachycardia, may be water-hammer in character; regular (mention all other points).
4. Face - Dusky and plethoric (due to polycythemia).
5. Hands - Warm and blue; may have flapping tremor.
6. Neck veins - Engorged and pulsatile (RVF).
7. Oedema - Bipedal pitting oedema (RVF).
8. Engorged veins may be seen in the forehead.
9. Nicotine staining of the finger (as commonly a smoker).

(C) Inspection :

1. Hyperactive accessory muscles of respiration with intercostal suction.
2. Barrel-shaped chest with features of emphysema.
3. Epigastric pulsation (RVH).

(D) Palpation :

1. Tracheal tug may be evident.
2. Apical impulse may not be localised due to emphysema (outward apex—RVH).
3. Palpable P₂ (diastolic shock).
4. Left parasternal heave (RVH).
5. Liver—Enlarged, soft and tender (RVF).

(E) Percussion :

Hyperresonant note all over the chest with loss of hepatic and cardiac dullness.

(F) Auscultation:

1. Diminished vesicular breath sound with prolonged expiration.
2. Occasional rhonchi with or without coarse crepitations.
3. Vocal resonance—Diminished all over the chest uniformly.
4. Loud P₂.
5. Right ventricular gallop (RVF) may be audible.
6. Pansystolic murmur of TI is audible in high pulmonary artery pressure.

Actually, we get a mixture of signs of emphysema PLUS signs of pulmonary hypertension. Emphysema overshadows all the inspeitory and palpatory signs in the chest, and we have to depend on **epigastric pulsation (RVH), engorged and pulsatile neck vein (RVF), and loud P₂ (due to pulmonary hypertension)** to diagnose a case of chronic cor pulmonale.

Why a COPD patient purses the lips ?

Pursing of lips during expiration helps the patient to maintain high intrabronchial pressure (i.e., to keep high end-expiratory pressure) above that within the surrounding alveoli (alveoli are overinflated or air-trapped in COPD) and thus prevents the collapse of bronchial walls by overinflated alveoli.

Who is a 'blue bloater' ?

In patients with 'predominant chronic bronchitis', there is severe hypoxia with hypercapnia, pulmonary hypertension and RVF from the very early stage though the ventilatory capacity is well preserved. They suffer from cyanosis (blue), oedema (bloated) and thus known as blue bloaters. Cough with sputum production is the main feature while dyspnoea is less common.

Who is a 'pink puffer' ?

In patients with 'predominant emphysema', there is gross impairment of ventilatory capacity and they usually suffer from severe disabling exertional dyspnoea. They are not cyanosed (so, pink) because the arterial O₂ is sufficient to saturate haemoglobin and try to blow off the excess CO₂ (puffer). They maintain a near-normal PaO₂ and PaCO₂. The patient is usually lean and thin.

Complications of COPD :

1. Recurrent pulmonary infections (very common).
2. Chronic cor pulmonale.
3. Spontaneous pneumothorax (due to rupture of emphysematous bulla).
4. Right ventricular failure (often a part and parcel of chronic cor pulmonale).
5. Type II respiratory failure.

What are the D/D you will entertain in your case ?

1. Chronic bronchitis —
 - a) Chronic cough with expectoration on most days for at least 3 consecutive months in a year, for more than 2 successive years.
 - b) Family history is absent.
 - c) Often a smoker.
 - d) Symptom-free childhood.
 - e) Episodes of frequent respiratory tract infection.
 - f) Copious mucoid sputum.
 - g) Wheeze and rhonchi+.

* Chronic bronchitis is associated with chronic cough with expectoration (phlegm). A patient of **chronic bronchitis with acute exacerbation** presents with pyrexia, and persistent or recurrent mucopurulent sputum production.

2. Tropical eosinophilia—

a) Nocturnal cough; breathlessness in a young patient.	c) Absolute eosinophil count > 3000/mm ³ .
b) H/O urticaria, worm infestation.	d) High level of filarial antibody.
	e) Miliary mottlings in the chest X-ray.
3. Allergic aspergillosis—

a) Episodic cough, wheeze and dyspnoea.	c) Sputum contains Aspergillus fumigatus.
b) Migratory densities in chest X-ray.	d) High eosinophil count in blood.
4. Cardiac asthma—Described in the section on 'Mitral stenosis'.

5. Acute bronchopneumonia—
 - a) No family history.
 - b) Extremes of ages.
 - c) Patient will be toxic.
 - d) Fever; bilateral patchy involvement affecting mainly the lower lobes.
 - e) Recently developing dyspnoea.

* D/D of 'acute severe asthma' in clinical practice are acute LVE, acute exacerbation of COPD, ARDS and tension pneumothorax.

** Recurrent bronchospasm is classically seen in bronchial asthma, recurrent pulmonary thromboembolism, tropical eosinophilia, carcinoid syndrome (tumour), acute exacerbation of chronic bronchitis/emphysema, and cardiac asthma (acute LVF).

Table 6 : Bedside differentiation between bronchial and cardiac asthma

Features	Bronchial asthma	Cardiac asthma
1. Past history-	Allergy and atopy; past H/O similar attacks	No H/O allergy: there may be few similar attacks in the past
2. Family history—	Often positive	Negative
3. Onset—	Late night or early morning	Early hours of night or midnight
4. Age—	Any age; usually young	Usually older age group
5. Clinical features-	<ol style="list-style-type: none"> a) Scanty, mucoid and tenacious expectoration b) Wheeze + + + c) Cyanosis + + d) Pulsus paradoxus e) BP—Usually normal f) Apex beat—Normal character g) There may be obliteration of liver and cardiac dullness h) RVH—May be present i) Heart sounds are muffled j) Rhonchi + + +, crepitations + k) May have 'silent chest' 	<ol style="list-style-type: none"> a) Profuse, pinkish and frothy expectoration b) Wheeze + c) Cyanosis + d) Pulsus alternans e) BP—May be hypertensive* f) Apex beat—Heaving or tapping g) Cardiac dullness may be increased due to LVH h) LVH—may be present i) Gallop rhythm present j) Rhonchi +, crepitations + + + k) Breath sound is not altered

* As hypertension is the commonest cause of cardiac asthma.

Complications of bronchial asthma :

1. Acute severe asthma.
2. Recurrent bronchitis and pneumonia (from infections).
3. Atelectasis (collapse) of lung or bronchiectasis (from mucous plugs).
4. Emphysema.
5. Pneumothorax (spontaneous).
6. **Complications of cough** like chest and abdominal wall soreness, exhaustion, severe vomiting, rib fracture (specially in elderly with osteoporosis or multiple myeloma), cough syncope, spontaneous pneumothorax, subconjunctival haemorrhage, frenal ulcer, urinary incontinence; prolapse of rectum, hernia or uterus.
7. Chronic cor pulmonale and respiratory failure (in late stage).

What is respiratory failure ?

It is a condition which is characterised by abnormality in blood gases ($\text{PaO}_2 < 60$ mm of Hg and / or $\text{PaCO}_2 > 50$ mm of Hg). The normal arterial blood gases are no longer maintained and the lung function is inadequate for the metabolic needs of the individual. It is of two types :

- 1 Type I — Low PaO_2 with normal or low PaCO_2 ; also known as hypoxaemic respiratory failure. It is found in ; pulmonary oedema (acute LVF), pneumonia, ARDS (acute lung injury), bronchial asthma, fibrosing alveolitis, pulmonary thromboembolism, pneumothorax, hysterical hyperventilation.

2. Type II — High PaCO_2 with low PaO_2 ; also known as hypercarbic failure or 'ventilatory failure'. It is found in : chronic bronchitis, emphysema, acute severe asthma, gross kyphoscoliosis, ankylosing spondylitis, trauma, neuromuscular disorders like myasthenia gravis, polyneuritis, poliomyelitis; depression of respiratory centre by narcotics or anaesthetics; end-stage of type I failure.

* **Commonest cause of respiratory failure is chronic bronchitis.**

Table 7 : Features of respiratory failure at the bedside

Features due to hypoxia :	Features due to hypercapnoea :
1. Restlessness, tachypnoea	1. Headache (bifrontal or occipital)
2. Impaired judgement	2. Reduced psychomotor activity
3. Mental confusion	3. Drowsiness, confusion or even coma
4. Depression of consciousness level	4. Flapping tremor (asterixis)
5. Central cyanosis	5. Water-hammer pulse and capillary pulsation
6. Tachycardia; systemic hypotension	6. Flushed and warm extremities (e.g., hands)
7. Cardiac arrhythmias	7. High blood pressure
8. Damage to organs like liver and kidney	8. Muscle twitching (fasciculations)
9. Convulsions	9. Papilloedema
10. Coma	10. Chemosis of the conjunctiva

Hypoxic features are more prominent in Type I failure. It was previously known as 'alcoholic type' as the patient becomes restless. Severe hypoxia may lead to bradycardia and systemic hypertension.

Features of hypercapnoea are more pronounced in Type II failure. It was previously known as 'an-aesthetic type' as the patient becomes drowsy. Majority of hypercapnoic features are due to cerebral and peripheral vasodilatation. This spectrum is also known as hypercapnoic encephalopathy or carbon dioxide narcosis.

* Features of **hypocapnoea** are : hyperventilation, dizziness, circumoral paraesthesia.

Bedside assessment of pulmonary function :

- | | |
|----------------------------|---|
| 1. Tachypnoea. | 7. Profuse sweating. |
| 2. Central cyanosis. | 8. Pulsus paradoxus. |
| 3. Rising pulse rate. | 9. Asynchronous respiration (i.e respiratory muscle fatigue). |
| 4. Inability to speak. | 10. Silent chest. |
| 5. Progressive exhaustion. | |
| 6. Altered sensorium. | |

If these features are present, immediately check the arterial blood gas and start mechanical ventilation accordingly.

What is ARDS ?

ARDS is a non-specific reaction of the lungs to a wide variety of insults, and by far the commonest one is sepsis. Previously called Adult Respiratory Distress Syndrome (ARDS) is now-a-days known as Acute Respiratory Distress Syndrome or '**acute lung injury**'. This is a severe form of type I respiratory failure caused by non-cardiogenic pulmonary oedema, or it is an acute hypoxaemic respiratory failure following a systemic or pulmonary insult without having any evidence of cardiac failure. Acute lung injury is a less severe disorder which can evolve into ARDS at any point of time. It is a serious disorder which is characterised by damage of alveolar as well as pulmonary capillary endothelium, where the alveoli become flooded with oedema fluid. The salient features are tachypnoea followed by dyspnoea, central cyanosis and crepitations throughout both lung fields; chest X-ray reveals diffuse, bilateral, interstitial and alveolar infiltrates ('fluffy' shadow). ARDS is also characterized by normal pulmonary capillary wedge pressure (<18 mm of Hg) and a $\text{PaO}_2/\text{F}_i\text{O}_2$ (inspiratory O_2 fraction) ratio < 200 mm of Hg. Read the causes of non-cardiogenic pulmonary oedema from the section on 'Mitral stenosis'.

How do you like to investigate bronchial asthma ?

1. Peripheral blood film — May show eosinophilia. Polymorphonuclear leucocytosis occurs in the presence of respiratory tract infection.
2. Sputum examination - Reveals,
 - a) Eosinophils,
 - b) Charcot—Leyden crystals (rhomboid crystals derived from eosinophils),
 - c) Laennec's pearls,
 - d) Curschmann's spiral (mucus casts of small airways),

- e) Dettrich's plugs, and
- f) Bronchial casts.
- 3. Chest X-ray - May be normal or show overinflation of lungs (due to emphysema). Rarely, there may be segmental or lobar collapse due to obstruction of bronchus by tenacious mucus. ***In worsening dyspnoea, always rule out the presence of pneumothorax.*** HRCT scan of thorax may be helpful to exclude other diseases.
- 4. Arterial blood gas analysis - In between the paroxysms, there is low PaO_2 with normal or low PaCO_2 . But in acute severe asthma, there is low PaO_2 with high PaCO_2 .
- 5. Lung function tests -
 - a) FEV₁.
 - b) FEV₁ / FVC.
 - c) PEF.

The effects are seen before and after application of bronchodilator drugs. These values are low in bronchial asthma. There is marked diurnal variation in PEF (no diurnal variation in chronic bronchitis or emphysema) and the value is lowest in the morning (early morning dipping).
- 6. Skin hypersensitivity tests - Done by 'prick method'. It may identify the allergen in an atopic individual (wheal and flare reaction after intradermal injection of common allergens).
- 7. Serum IgE — Elevated serum IgE level supports atopy.

What will be the outline of management of bronchial asthma ?

1. Rest in bed in propped-up position with moist O_2 inhalation at the rate of 4-6 litre / minute.
2. Salbutamol or terbutaline—I.M or I.V route in a dose of 250-500 μg , or inj. aminophylline with a loading dose of 5.0 mg/kg of body weight by slow I.V infusion over 20 minutes and maintenance dose of 0.5 mg/kg/hour, given as a continuous IV infusion (in acute case). Tablet salbutamol (2 mg), terbutaline (2.5 mg) or theophylline (100 mg) may be given three times daily in between the attacks. Long-acting (β_2 -stimulants like salmeterol or formoterol are now used routinely as inhaler or spacer.
3. Corticosteroid—Hydrocortisone hemisuccinate 200 mg I.V may be given in acute severe asthma as a loading dose, followed several hours later by an infusion regulated to deliver 3 mg/kg every 6 hours. Now-a-days corticosteroid (as inhaler) is also used in between the attacks.
4. Sodium chromoglycate inhalation 20 mg, four times daily in between the attacks; recently nedocromil sodium is used as 4 mg, 2-4 times daily.
5. Ipratropium bromide (anticholinergic) inhalation, or tablet ketotifen (antihistaminic) 1 mg twice daily may be given in between the attacks.
6. Leucotriene antagonist, montelukast (10 mg, orally, once daily) or zafirlukast (20 mg, orally, BD) may be used to provide control of mild to moderate persistent asthma.
7. Sedatives, tranquillisers or narcotics should preferably be avoided.
8. Treatment of pulmonary infection with proper antibiotics. Expectorants or mucolytics (bromhexine, acetyl-cysteine) may be used.
9. Desensitisation or immunotherapy by repeated subcutaneous injections of gradually increasing dose of extracts of allergen/allergens may be tried (doubtful value).
10. Recent development—Omalizumab (recombinant humanized monoclonal antibody which modulates IgE-associated inflammation) or etanercept (anti-TNF therapy) may be tried in corticosteroid-resistant severe asthma.

* Acute severe asthma needs special management. Read the step-up and step-down drug treatment of bronchial asthma from any standard text book.

Side effects of pure (100%) oxygen therapy :

1. Cough and bronchial irritation as it is both irritant and toxic; drying of secretions.
2. Carbon dioxide narcosis occurs when high O_2 concentration is given to patients with respiratory failure who thrives on 'hypoxic drive', e.g., in chronic bronchitis'.
3. Retrolental fibroplasia (blindness) and bronchopulmonary dysplasia—in premature infants.
4. Pulmonary oedema and ARDS.
5. Consolidation of the lung.
6. Absorption collapse (in low ventilation/perfusion zones).
7. Respiratory depression.

* Newborn and infants are more susceptible to effects of pure O_2 than older children and adults.

Conclusion :

The **hallmark of diagnosis** of bronchial asthma is sibilant, polyphonic rhonchi (mainly expiratory) present all over the chest.

----- O -----

Section 3

GASTROINTESTINAL SYSTEM

The cardinal symptoms of G.I. system are enlisted below :

1. Anorexia (loss of appetite), dyspepsia, indigestion
2. Dysphagia, odynophagia (painful swallowing); toothache, bleeding gum, dentures
3. Nausea, vomiting, retching, eructation, hiccup (singultus)
4. Heartburn (pyrosis i.e., a hot, burning retrosternal discomfort), water brash (salivary hypersecretion) and regurgitation (effortless appearance of gastric contents in mouth)
5. Jaundice, yellow urine, pruritus
6. Pain abdomen (including biliary colic)
7. Haematemesis
8. Melaena
9. Haematochezia
10. Bleeding per rectum
11. Alteration in bowel habit
12. Diarrhoea or constipation, mucus in stool, worms in stool.
13. Swelling in the abdomen, distension of the abdomen or mass in the abdomen.
14. Wind or flatulence (belching or flatus)
15. Loss of weight
16. Pedal oedema
17. Oliguria (e.g., hepato-renal syndrome from cirrhosis of liver)
18. Neuropsychiatric manifestations (inversion of sleep rhythm, drowsiness from hepatic pre-coma)
19. Fever (intestinal tuberculosis, lymphoma, spontaneous bacterial peritonitis)
20. General symptoms—fatigue, bladder symptoms etc.

* **Dyspepsia** is the summation of a variety of G.I. symptoms e.g., anorexia, early satiety plus flatulence.

** Difficulty in defecation with straining (dyschezia), and feeling of incomplete evacuation with a constant desire for defecation (tenesmus) fall within 'alteration in bowel habit'. Rule out malignancy of bowel in recent alteration in bowel habit.

*** Don't forget to take the H/O hernia or haemorrhoids. Other minor symptoms are halitosis, dry mouth, ulcers in mouth (e.g., aphthous ulcer) or pain in the oral cavity.

**** Too much flatus production is commonly due to aerophagia (air ingestion), malabsorption or lactase deficiency.

***** Terminology associated with alimentary system :

- Appetite—pleasurable desire to eat
- Hunger—unpleasant sensation produced as a result of empty stomach as well as peristaltic contraction
- Satiety—a sensation of satisfaction after taking food
- Sitophobia—fear of eating (e.g., in oropharyngeal dysphagia, pulmonary aspiration of food occurs during swallowing)
- Phagophobia—fear of swallowing (e.g., in rabies, tetanus and hysteria)
- Globus pharyngeus—sensation of something lodged in the throat or lump in the throat
- Nausea—feeling or desire to vomit
- Retching—laboured and rhythmic contraction of respiratory and abdominal muscles preceding vomiting
- Heartburn (pyrosis)—burning sensation or a sensation of warmth located substernally
- Dysgeusia—altered taste sensation

Scheme of Examination

(A) UPPER G.I. TRACT :

- | | |
|---------------------------|-------------------------------|
| 1. Lips. | 6. Fauces and tonsils. |
| 2. Teeth. | 7. Palate. |
| 3. Gum. | 8. Breath (halitosis or foul) |
| 4. Cheek (buccal mucosa). | 9. Oropharynx. |
| 5. Tongue. | |

* A torch, a tongue depressor and a pair of gloves are necessary for examination of oral cavity. It is not possible to examine the oesophagus at the bedside; ask the patient for dysphagia, odynophagia, heart burn, or give the patient a glass of water or biscuits, and ask him to swallow. Oesophageal disorder is analysed by barium swallow, upper G.I endoscopy, and motility study by manometry and isotope Tc" taken with solid or liquid food.

(B) EXAMINATION OF THE ABDOMEN :

I. INSPECTION :

1. Shape of the abdomen.
2. Flanks - Full or not (presence of free fluid within the peritoneal cavity i.e., ascites makes the flanks 'full').
3. Venous prominence (examine in sitting or standing position with asking the patient to cough).
4. Umbilicus -
 - (i) Position of the umbilicus and the slit :
 - a) Whether inverted or everted.
 - b) Whether situated midway between xiphisternum and symphysis pubis, or not.
 - c) Whether the slit is circular, transverse or vertical.
 - (ii) Venous prominence around the umbilicus (caput medusae from portal hypertension).
 - (iii) Hernia (umbilical).
 - (iv) Granuloma, adenoma (raspberry tumour), endometriosis (looks raw and red).
 - (v) Nodules around the umbilicus (e.g., metastatic nodule from intra-abdominal malignancy).
 - (vi) Bluish discolouration around the umbilicus (Cullen's sign from haemoperitoneum) or Grey Turner's sign (discolouration of skin in the abdominal flanks due to haemoperitoneum).
 - (vii) Omphalolith (inspissated desquamated epithelium with other debris)—specially in elderly.
 - (viii) Sinus (malignant or tuberculous peritonitis, Crohn's disease, patent urachus).
5. Condition of the skin — Ulcer, striae, pigmentation, scar and puncture mark, sinus, keloid.
6. Any localised swelling.
7. Movement of the abdomen —
 - (i) Respiratory movement.
 - (ii) Pulsation.
 - (iii) Peristalsis.
8. Hernial sites like —
 - (i) Inguinal,
 - (ii) Femoral,
 - (iii) Umbilical,
 - (iv) Epigastric, or
 - (v) Incisional hernia.
9. Hair (secondary sexual hair—Loss of pubic hair, virilism etc).

* During inspection of the abdomen, extend the legs of the patient.

** Ask the patient to stand and note the presence of impulse on coughing on the hernial swellings.

*** For observation of respiratory movements, pulsation and peristalsis—squat down by the side of the bed to have a tangential inspection.

**** Finally, inspect the posterior abdominal wall (back), groins, penis and scrotum.

II. PALPATION :

- a) Superficial —

1. Surface temperature.
2. Tenderness or hyperaesthesia.
3. Parietal oedema.
4. Consistency or feel (normal elastic feel, or muscle guard or rigidity).
5. Localised lump.
6. Divarication of recti (if any) by rising test.
7. Direction of blood flow in prominent veins.
8. Pulsation (transmitted or expansile).
9. Fluid thrill with girth of the abdomen at the level of the umbilicus.

* Measure the girth of the abdomen at the level of umbilicus with the help of a tape.

** Divarication of recti—two rectus abdominis muscles are widely separated.

b) Deep —

1. Liver (in details).
2. Spleen (in details).
3. Gall bladder.
4. Kidneys (in details).
5. Colon.
6. Palpation of the testes - It is a must.
7. Any other lump in the abdomen.
8. Deep tender spots - McBurney's point, gall bladder point, epigastric point, renal angle etc.
9. Rebound tenderness.
10. Examination of hernia and external genitalia (for examination of hernia, ask the patient to stand and cough with turning the head to other side). Palpate both groins.
11. The aorta, paraaortic glands and common femoral vessels.
12. Urinary bladder.

* **Rebound tenderness**—First give firm pressure over the abdomen, and then 'suddenly' take off the hands. The patient complains of severe pain in the abdomen if the sign is positive. This is positive in peritonitis (basically inflammation of perietal peritoneum) from any cause, including acute appendicitis.

** For deep palpation, conventionally palpate with the dominant hand. In any obese, muscular or poorly relaxed patient, the left hand is placed over the right hand to exert more pressure during palpation. Dipping method is adopted in the presence of ascites. The **abdominal aorta** is palpated by placing both hands vertically with extended fingers (eight fingers of both hands except thumbs), which are held side by side, and the palpation is carried a little above and to the left of umbilicus.

*** Groin should be carefully examined for inguinal lymph nodes, hernia and femoral pulsation.

**** Enquire about pain in the abdomen and it is preferable to examine that part at the last.

III. PERCUSSION :

1. General note of the abdomen (tympanitic note in health is elicited by 'light' percussion).
2. Upper border of liver (and splenic) dullness.
3. Band of colonic resonance over a kidney lump.
4. Shifting dullness.
5. Urinary bladder - Full or empty.

IV. AUSCULTATION :

1. Peristalsis.
2. Hepatic / splenic rub.
3. Venous hum.
4. Bruit (e.g., hepatic bruit, renal artery bruit).
5. Ausculto-percussion and succussion splash (in pyloric stenosis).

(C) PER RECTAL EXAMINATION (optional) :

* Abdominal examination is never completed without examining external genitalia and performing rectal examination. Per rectal examination is important in females to note fullness of pouch of Douglas by ovarian cyst, fluid collection or malignant deposits. Per vaginal examination is often helpful to diagnose the nature of an abdominal mass.

Case 13

PYLORIC STENOSIS

What is your diagnosis ?

This is a patient of pyloric stenosis possibly developed from chronic duodenal ulcer.

Reasons behind your diagnosis :

This is a case of pyloric stenosis because :

1. The patient is giving a long history of duodenal ulcer. Previously the pain was present in epigastrium, appeared in empty stomach (or there was H/O hunger pain) with history of heartburn, water brash, nocturnal pain and pointing sign. The pain was relieved by taking food or antacid.
2. Distension of the upper part of abdomen with foul smelling gaseous eructation and belching.
3. Nausea as well as projectile vomiting is present which contains food taken by the patient 1-2 days back and is copious in amount, and the smell of the vomitus is very offensive. Vomiting greatly relieves the patient. Vomiting is often self-induced.
4. The typical character of epigastric pain (of chronic duodenal ulcer) is now changed and instead a sense of epigastric fullness or discomfort is present. The periodicity of pain is lost. The pain has lost its relationship with food in the last few months.
5. Rapid loss of weight and increasing constipation.
6. Features of tetany is present (alkalosis caused by vomiting) - Say, if present; signs of dehydration are present.
7. **A diffuse swelling** is present in the upper abdomen.
8. **Visible peristalsis** from left to right hypochondrium is seen (see the section on 'Visible peristalsis').
9. **Succussion splash** is heard (splashing sound heard on shaking the patient's abdomen).
10. **Ausculto-percussion** over the abdomen reveals gastric distension.

* A lump in the upper abdomen may be felt in case of carcinoma of the pylorus presenting as gastric outlet obstruction.

What is your case ?

Build up the summary from the history and physical findings.

Aetiology of 'gastric outlet obstruction' :

- | | |
|---|---|
| 1. Chronic duodenal ulcer. | 6. Hypertrophic gastritis, rarely. |
| 2. Carcinoma of the stomach (antrum). | 7. Gastroparesis (e.g., from diabetes mellitus) |
| 3. Oedema of the pylorus. | 8. Bezoars (trichobezoars or phytobezoars). |
| 4. Adult hypertrophic pyloric stenosis. | 9. Gastric lymphoma. |
| 5. Congenital hypertrophic pyloric stenosis (in infants). | |

* Chronic duodenal ulcer -> duodenal cicatrization -> fibrotic stricture -> pyloric stenosis.

Common differential diagnosis you will consider in your case :

1. Carcinoma of the antrum :
 - a) Age will be higher.
 - b) History is short.
 - c) Classical presentation of gastric carcinoma (vide infra).
 - d) Rarely, epigastric pain relieves with food.
2. Adult hypertrophic pyloric stenosis :
 - a) No past H/O peptic ulcer disease.
 - b) Other features are like classical pyloric stenosis.
3. Intestinal obstruction :
 - a) Peristalsis will be periumbilical.
 - b) Abdomen will be tympanitic.
 - c) Obstipation (absolute constipation, i.e., no passage of faeces and flatus).
 - d) Bowel sounds gradually diminish.
4. Primary carcinoma of liver (predominantly involving the left lobe) :
 - a) History is short.

- b) Jaundice (rare).
- c) Vomitus is not offensive.
- d) Neither peristalsis seen nor any succussion splash heard.
- e) Hard, tender, nodular liver; emaciation.

Common physical signs in gastric carcinoma :

1. Tender mass in the epigastrium (hard lump).
2. **Anaemia.**
3. **Asthenia** (cachexia).
4. Jaundice (due to hepatic metastasis).
5. Liver - Enlarged, tender, hard with nodular surface and sharp margin; hepatic rub may be present due to metastasis.
6. Palpable Virchow's gland (palpable hard and fixed lymph nodes in between two heads of left stemomastoid muscle - **Troisier's sign**).
7. Metastatic nodules around the umbilicus (Sister Marie Joseph's nodule).*
8. Ascites (due to malignant peritonitis).
9. Visible gastric peristalsis.
10. Succussion splash (due to gastric outlet obstruction).
11. Krukenberg's tumour (due to metastasis in the ovary) - Detected by P/V examination.
12. Blumer's shelf (shelf-like mass in the prerectal pouch) - Detected by P/R examination.
13. Rarely, there may be dermatological features like acanthosis nigricans, dermatomyositis, migratory thrombophlebitis (Trousseau's sign) etc.

N.B. : Examination of neck glands, jaundice, anaemia, ascites are important in a patient suffering from gastric outlet obstruction. **Anorexia** is a very important symptom. The tympanicity of Traube's space percussion is often lost.

* The Sister was nursing superintendant of St. Mary's Hospital, Rochester. USA.

How to elicit succussion splash (Hippocratic succussion) ?

This is the characteristic splashing sound heard in pyloric obstruction when gastric contents are retained for more than 4 hours in the stomach. The sign is elicited by shaking the patient from side to side and simultaneously listening the splashing sound by placing the diaphragm of stethoscope over the distended area in the epigastrium.

The splash is often realised by palpation (by placing hands over epigastrium) and heard without the help of stethoscope. This splashing sound may be normally present after taking a large amount of water.

How to elicit ausculto-percussion ?

This method helps to **delineate the enlarged stomach** in cases of pyloric stenosis and sometimes in acute dilatation of the stomach. Put the diaphragm of stethoscope over the epigastrium and scratch the upper part of the abdomen by finger or a pen/ pencil in a centrifugal fashion downwards and sideways. Note the sudden change in character of the sound and mark individual points with a skin pencil. The pencil marks are joined to get the outline of distended stomach which generally crosses the umbilicus in a patient with pyloric stenosis. This method is called 'ausculto-scratching'. In the classical method of ausculto-percussion, a doctor percusses the stomach centrifugally (in the direction away from the stethoscope) while the other doctor listens the sudden changes in character of the sound.

Medical causes of peptic ulcer :

- | | |
|-------------------------|-------------------------------------|
| 1. Cirrhosis of liver. | 5. Cushing's syndrome. |
| 2. COPD. | 6. Chronic renal failure. |
| 3. Hyperparathyroidism. | 7. Systemic mastocytosis. |
| 4. Polycythemia vera. | 8. NSAID or corticosteroid therapy. |

How will you confirm your diagnosis in a case of gastric outlet obstruction ?

1. Typical history with physical findings.
2. Analysis of gastric contents (fractional test meal) —
 - a) Volume of gastric aspiration will be > 200 ml after overnight fasting.
 - b) The colour of the juice is coffee-ground (in gastric carcinoma).
 - c) Bile is absent in all the samples.
 - d) Aspirate is offensive and contains food residue (classical pyloric stenosis).
 - e) Pentagastrin-fast achlorhydria virtually diagnoses gastric malignancy.

3. Barium meal X-ray of stomach and duodenum -
 - a) Grossly dilated stomach with excessive fasting contents.
 - b) Lesion at or near the pylorus.
 - c) **Failure of the stomach to evacuate the meal even after 6 hours (characteristic).**
4. Upper G. I. endoscopy - Diagnostic and a standard practice now.
 - a) Can identify the obstruction and its degree.
 - b) Old food material may be seen.
 - c) May identify carcinoma in the antrum (biopsy may be taken).
5. FNAC (fine needle aspiration cytology) or excision biopsy of a palpable cervical lymph node (may diagnose carcinoma of stomach causing gastric outlet obstruction).
6. Liver biopsy may be done (to diagnose metastasis from gastric carcinoma).
7. Blood biochemistry - Persistent vomiting may give rise to hypokalaemia, hypochloraemia and alkalosis. Hyponatraemia and high blood urea level may be present.

* Coffee-ground colour vomitus (gastric carcinoma) is due to conversion of haemoglobin to acid haematin by gastric acid.

How are you going to manage your case of pyloric stenosis ?

1. Repeated stomach wash or gastric decompression is done (at 2-4 hourly interval for 3-4 days) to remove all food debris.
2. Correction of dehydration, dyselectrolytaemia and metabolic alkalosis are done by :
 - a) 5% dextrose and normal saline.
 - b) Injection potassium chloride 80-100 mmol / day.
3. Proton pump inhibitors may reduce pyloric oedema or heal ulcer.
4. Gradually the patient is put on oral diet. At first liquid, then semisolid diet is started.
5. Elective surgery—Surgery is required in majority of patients in the long run. Vagotomy with gastrojejunostomy, or subtotal gastrectomy is done according to the choice of the surgeon. Balloon dilatation of the obstructed outlet may be done by upper G.I. endoscopy.

Late complications after gastric surgery :

1. Recurrent ulceration.
2. Afferent loop syndrome.
3. Dumping syndrome—Early and late.
4. Bile reflux gastropathy.
5. Postvagotomy diarrhoea.
6. Osteomalacia and osteoporosis.
7. Haematological complications—Iron deficiency anaemia, megaloblastic anaemia
8. Maldigestion and malabsorption.
9. Small stomach syndrome.
10. Adenocarcinoma of gastric stump.

Describe the normal peristaltic sound :

'Bowel sounds' are produced by the movement of faeces, fluid and flatus within the bowel lumen as a result of peristalsis. Place the stethoscope on one side of the abdominal wall and keep it there minimally for one minute until the bowel sounds are heard. Usually the stethoscope is placed just to the right of the umbilicus. Stethoscope should not be shifted from site to site. Normal bowel sounds are intermittent (at 5 or 10-second's interval) low-or medium-pitched gurgles mixed with occasional high-pitched tinkle.

In mechanical intestinal obstruction—Frequent loud low-pitched gurgles (borborygmi) are heard, often interspersed with high-pitched tinkles occurring in a rhythmic pattern with peristalsis. As a whole, the peristaltic sounds are **exaggerated**.

In paralytic ileus and peritonitis—Abdomen is **silent**.

N.B. : Bowel sounds are also exaggerated in malabsorption and severe G.I. bleeding.

* While dealing with a patient related to gastrointestinal system, one should be careful in examining :

- | | |
|---|---|
| 1. Anaemia. | 6. Hernial sites. |
| 2. Jaundice. | 7. Testes. |
| 3. Neck glands (Virchow's gland specially). | 8. Spine (referred pain from caries spine). |
| 4. Oedema (nutritional, malabsorption). | 9. Rectum (per rectal examination), and |
| 5. Ascites (cirrhosis, peritonitis). | 10. Auscultation of peristaltic sounds. |

Case 14

ACUTE VIRAL HEPATITIS

What is your diagnosis?

This is a case of acute viral hepatitis probably due to type B virus infection, the patient is in the icteric stage and without any sign of hepato-cellular failure at present.

What is the reason behind your diagnosis?

This is a case of acute viral hepatitis because :

The patient suffered from high fever with mild chill and rigor, aches and pain all over the body 8 days back, and these ailments persisted for next 5 days. Initially, it was associated with profound anorexia, nausea and vomiting. At the onset, he also had urticarial rashes and arthralgia. The colour of the urine was yellow from the very beginning, and the patient complained of yellowish discolouration of eyes for last 5 days. The patient is having slightly pale stool from day before yesterday. He also complained of dull-aching, non-colic, non-radiating pain in the right hypochondrium. Neither he is suffering from generalised pruritus nor any bleeding diathesis at present. There is no past H/O dental extractions, tattooing, acupuncture, blood taken (by unsterile syringe) for any investigational purpose within last 6 months or 'high risk behaviour' (all predispose to type B infection), jaundice, biliary colic, drug intake (like OC pills, rifampicin, INH), amoebic dysentery or alcohol intake. There was H/O receiving blood transfusion 3 months back when he suffered from a serious street accident. No family member or persons in his locality is suffering from similar type of illness (vide Type A infection). Thus, the disease started acutely and is progressing gradually. For the last 8 days of illness, he is feeling extremely weak.

On examination, there is presence of moderate degree of jaundice; liver is enlarged 4 cm below the right costal margin at right MCL, soft, tender with rounded and regular margin; it has a smooth surface and there is absence of pulsation, rub or bruit. Spleen is just palpable, soft and non-tender. The gall bladder is not palpable and there is discomfort on percussion over the right lower chest. There is neither any free fluid in the abdomen nor any sign of hepato-cellular failure present. CVS, nervous system and respiratory systems are essentially within normal limit.

* Splenomegaly and cervical adenopathy are present in 10-20% cases.

Give the summary of the case :

Described above.

'Extrahepatic associations' of type B infection :

1. Serum sickness-like' syndrome manifested by arthralgia or arthritis, fever, urticarial rash (present in this patient), angio-oedema and rarely haematuria or proteinuria.
2. Polyarteritis nodosa (systemic necrotising vasculitis).
3. Acute glomerulonephritis.
4. Polymyalgia rheumatica.
5. Essential mixed cryoglobulinaemia (arthritis with cutaneous vasculitis; commoner in hepatitis C).
6. Guillain-Barre syndrome.
7. Myocarditis.

* Porphyria cutanea tarda, essential mixed cryoglobulinaemia, lichen planus and sicca syndrome are clinical associations with hepatitis C.

Chief complaints of your patient :

1. Anorexia, nausea and vomiting for last 8 days.
2. Yellowish discolouration of urine for last 7 days.
3. Yellowish discolouration of eyes for last 5 days.
4. Fever for 5 days (initial 5 days of illness).
5. Pain in the right upper abdomen for last 3 days.

Common symptoms of acute viral hepatitis :

1. Anorexia, nausea and vomiting.
2. Arthralgia, myalgia and headache.
3. Pharyngitis, cough and coryza.
4. Fatigue and malaise.

5. Dark urine.
6. Clay-coloured stool (due to canalicular obstruction caused by swelling of hepatocytes).

Probable causes **of marked anorexia** :

1. Acute viral hepatitis.
2. Tuberculosis.
3. Malignancies, specially carcinoma of the stomach (TNF- α is an anorectic substance).
4. Chronic diseases like cirrhosis of liver, chronic renal failure, CCF.
5. Drugs e.g., chloroquine, quinine, digitalis, metronidazole, erythromycin.
6. Psychogenic—depression, emotional upset, anorexia nervosa.
7. Chronic smoking, chronic alcoholism.

Agents responsible for acute viral hepatitis :

- | | |
|--------------------------------|--------------------------|
| 1. Hepatitis A, B, C, D and E. | 4. Cytomegalovirus. |
| 2. Yellow fever virus. | 5. Herpes simplex virus. |
| 3. Epstein-Barr virus. | 6. Coxsackie B virus. |

* Other infective causes of acute hepatitis are : non-viral—leptospira (Weil's syndrome), toxoplasma and coxiella; post-viral—Reye's syndrome in children (aspirin-induced).

Features of different types of viral hepatitis (A to E) :

Type A :

1. Transmission—Faecal-oral route commonly (percutaneous and sexual route—uncommon).
2. Age—Children and young adults.
3. Seasonal influence—In winter and rainy seasons.
4. Incubation period—15 days to 45 days (mean 30 days).
5. Severity—Generally mild.
6. Chronicity—Never occurs.
7. Prognosis—Excellent.
8. Carrier state—Never occurs.
9. Australia antigen (HB_sAg)—Negative.
10. Peak ALT—800 to 1000 IU/L.

Type B :

1. Transmission—Usually parenteral (also sexually transmitted).
2. Age—Any age group may be affected (young adults, babies, toddlers).
3. Seasonal influence—Occurs round the year.
4. Incubation period—30 days to 180 days (mean 60-90 days).
5. Severity—Occasionally severe.
6. Chronicity—Occurs in 1-10% (90% of neonates).
7. Prognosis—Not bad but worse with increasing age.
8. Carrier state—Occurs (1-30%).
9. Australia antigen—Positive.
10. Peak ALT—1000 to 1500 IU/L.

* Humans are the only source of infection. HB_sAb is protective.

Type C :

1. Transmission—Parenteral (sexual transmission uncommon).
2. Age—Any age may be affected (common in adults).
3. Seasonal influence—Occurs round the year.
4. Incubation period—15 days to 160 days (mean 50 days).
5. Severity—Moderate.
6. Chronicity—Commonly occurs (85%).
7. Prognosis—Not good, not bad (moderate).
8. Carrier state—Exists (0.5-1%).
9. Australia antigen—Negative.
10. Peak ALT—300 to 800 IU/L.

* Accounts for more than 90% of post-transfusion hepatitis. Anti-HCV is not protective.

Type D (Delta virus) :

1. Transmission—Commonly parenteral (also sexually transmitted).
2. Age—Any age (as type B virus).
3. Seasonal influence—Occurs round the year (as type B virus).
4. Incubation period—30 days to 180 days (mean 60-90 days).
5. Severity—Occasionally severe.
6. Chronicity—Commonly occurs.
7. Prognosis—Good in acute variety, poor in chronic variety.
8. Carrier state—Variable (10-20% in pregnant women).
9. Australia antigen—Positive (as there is 'coinfection' with or 'superinfection' on type B).
10. Peak ALT—1000 to 1500 IU/L.

* Delta virus requires hepatitis B virus for replication i.e., it has no independent existence.

Type E :

1. Transmission—Only by faecal-oral route.
2. Age—Young adults.
3. Seasonal influence—Commonly after monsoon and flood but sporadic, isolated cases occur.
4. Incubation period—15 days to 60 days (mean 40 days).
5. Severity—Usually mild.
6. Chronicity—None.
7. Prognosis—Good (mortality is very high in women in the last trimester of pregnancy).
8. Carrier state—Does not exist.
9. Australia antigen—Negative.
10. Peak ALT—800 to 1000 IU/L.

* Faecal-oral route—Type A and E; Parenteral route—Type B, C, D.

Chronicity—Type B, C, D.

** Hepatitis C is the commonest cause of post-transfusion hepatitis.

*** Hepatitis A, B and C viruses are transmitted through saliva.

**** Type A and Type B hepatitis were previously known as infective hepatitis and serum hepatitis respectively. Hepatitis C and hepatitis D are also known as non-A non-B hepatitis and delta virus hepatitis respectively.

Describe hepatitis G virus (HGV) :

It is a RNA virus (hepatitis A, C, D, E and G are RNA virus; hepatitis B virus is a DNA virus). HGV is transmitted through blood transfusion (risk factors are like HCV). It is the sixth heterotropic viral agent. Extensive investigations have failed to show that HGV plays a causal role in acute or chronic liver disease and thus, it does not seem to be a serious human pathogen. HGV coinfection may improve survival in HIV infected patients.

Hepatitis TT virus (initials of name of a Japanese patient; DNA virus) is also unlikely to be a significant cause of liver diseases.

Type of jaundice in your case :

It is hepato-cellular jaundice.

Complications of acute viral hepatitis :

1. 'Serum sickness-like' syndrome (described previously).
2. Fulminant hepatitis (massive hepatic necrosis)—the most feared complication.
3. Cholestatic viral hepatitis (protracted cholestatic jaundice).
4. Relapsing hepatitis (biochemical/clinical)—return of symptoms and signs of acute hepatitis during recovery phase.
5. Post-hepatitis syndrome (symptoms persist though LFT remains more or less normal).
6. Chronic carrier state.
7. Chronic hepatitis (lobular, persistent and active).
8. Cirrhosis of liver (postnecrotic variety).
9. Hepato-cellular carcinoma.
10. Rare—Pancreatitis, myocarditis, aplastic anaemia, peripheral neuropathy, atypical pneumonia, transverse myelitis, papular acrodermatitis in children, renal failure, polyarteritis nodosa.

Stages of acute viral hepatitis :

There are three stages :

1. Prodromal stage,
2. Icteric stage (i.e., stage of clinical jaundice), and
3. Recovery stage.

Clinical types of viral hepatitis :

- | | |
|---|-----------------------------|
| 1. Acute icteric hepatitis—common type. | 5. Cholestatic hepatitis. |
| 2. Anicteric hepatitis. | 6. Post-hepatitis syndrome. |
| 3. Fulminant hepatitis. | 7. Chronic hepatitis. |
| 4. Relapsing hepatitis. | |

What is anicteric hepatitis ?

Here, the jaundice is not clinically evident, and symptoms are few though the liver becomes enlarged and tender. It is a mild illness though there is bilirubinuria and elevation of aminotransferases. These patients usually suffer from chronic liver disease (e.g., chronic active hepatitis) later in life. The diagnosis is difficult and requires a high index of suspicion.

Differential diagnosis you will consider in this case :

1. Differentiate among the 5 types of viral hepatitis (the present case suffers from type B infection. There is H/O receiving blood transfusion and urticaria, arthralgia at the onset of the disease).
2. Hepatic amoebiasis or amoebic liver abscess —
 - a) H/O amoebic dysentery may be present.
 - b) Liver is enlarged and very tender.
 - c) Intercostal tenderness is present.
 - d) Fever with chill and rigor, excessive sweating at night.
 - e) Patient looks toxic.
 - f) Jaundice, splenomegaly, marked anorexia are not very common features.
3. Alcoholic hepatitis—
 - a) H/O chronic alcoholism.
 - b) Stigmata of alcoholic liver disease (read the section on 'Cirrhosis of liver').
 - c) Jaundice with enlarged, tender liver.
 - d) Anorexia, weight loss.
4. Carcinoma of liver—
 - a) Commonly affects middle aged or older persons.
 - b) Progressive jaundice may be present (commonly in secondary malignancy of liver).
 - c) Liver feels hard, tender and nodular.
 - d) Anaemia, hard lymph nodes in neck and cachexia are prominent features.
 - e) Fever and splenomegaly are usually absent.
 - f) Ascites is not uncommon.
5. Drug-induced hepatitis—
 - a) H/O intake of hepatotoxic drugs (rifampicin, INH, oral contraceptive pills).
 - b) Malaise before onset of jaundice.
 - c) Occasionally arthralgia, rash and fever are present.
 - d) Liver is enlarged and may be tender.

* It is often very difficult to differentiate this entity from acute viral hepatitis.
6. Haemolytic jaundice—
 - a) Anaemia with mild jaundice.
 - b) Freshly voided urine is acholuric (not yellow).
 - c) Leg ulcers may be present.
 - d) Typical fates may be revealed (vide Thalassaemia).
 - e) Splenomegaly in almost all the cases.
 - f) Non-tender liver, if at all palpable.
7. Congestive cardiac failure—
 - a) Cardiomegaly with underlying heart disease.

- b) Features of CCF.
- e) Liver is enlarged, soft and tender.
- d) Clinical jaundice is not commonly seen (latent jaundice may be present).
- 8. Acute malaria—
 - a) Fever with chill and rigor. Typical paroxysm with cold, hot and sweating stage.
 - b) Anaemia.
 - c) Anorexia and jaundice are not so common.
 - d) Splenomegaly is more prominent than hepatomegaly.
- 9. Cirrhosis of liver—
 - a) Long H/O dyspeptic symptoms.
 - b) H/O G.I. bleeding.
 - c) Signs of portal hypertension and hepato-cellular failure.
 - d) Liver is enlarged but non-tender.
 - e) Splenomegaly.
- 10. Chronic hepatitis (see below).
- 11. Yellow fever, Weil's disease, infectious mononucleosis, obstructive jaundice etc.

Enumerate the causes of 'prolonged jaundice' :

Jaundice present for more than 6 months may be arbitrarily called as 'prolonged jaundice'. The common causes are,

- 1. Cholestatic viral hepatitis.
- 2. Chronic hepatitis.
- 3. Cirrhosis of liver.
- 4. Carcinoma of liver (secondary commonly).
- 5. Thalassaemia.
- 6. Drug-induced hepatitis (e.g., rifampicin, INH, chlorpromazine).
- 7. Extrahepatic biliary obstruction (e.g., stone, stricture, carcinoma of the head of pancreas).
- 8. Alcoholic hepatitis.
- 9. Wilson's disease.
- 10. Other causes like Gilbert's syndrome, primary biliary cirrhosis, hereditary spherocytosis, sickle cell anaemia, autoimmune haemolytic anaemia, sclerosing cholangitis etc.

* 'Prolonged jaundice' may be given as a long **case**.

Causes of enlarged and tender liver :

Read the section on 'Hepatosplenomegaly'.

What is cholestatic viral hepatitis ?

It is commonly associated with type A (more common) or type B infection. The jaundice gradually deepens and intense pruritus appears. Jaundice, hepatomegaly and LFT abnormalities may persist for many months but ultimately the recovery is complete (i.e., prognosis is good). It is often very difficult to differentiate this condition from surgical cholestasis (read the section on 'Jaundice').

Give an out line of chronic hepatitis ?

(A) Definition—

It is a chronic inflammatory reaction in the liver present on clinical or other grounds (biochemical/serological) without improvement for more than 6 months.

(B) Aetiology—

- 1. Autoimmune hepatitis.
- 2. Hepatitis B, C, and D.
- 3. Drugs (methyl dopa, INH, oxyphenisatin, ketoconazole, nitrofurantoin).
- 4. Wilson's disease, α_1 -antitrypsin deficiency, alcoholic liver disease, non-alcoholic steatohepatitis (NASH) and ulcerative colitis.
- 5) Cryptogenic.

(C) Types—

- 1. Chronic lobular hepatitis—Histology resembles unresolved acute viral hepatitis.

2. Chronic persistent hepatitis—The limiting plate between hepatocytes and portal zones is intact, and piecemeal necrosis is not seen. There is expansion of the portal zone by infiltration with mononuclear cells.
3. Chronic active hepatitis (severe form)—There is erosion of limiting plate, presence of piecemeal necrosis, formation of 'rosette' and 'bridging necrosis'; prognosis of this disease is very bad.

* No. 1 and 2 are milder forms.

** The above described 3 categories are not very helpful for prognostication. The new classification of chronic hepatitis is based on 1. Aetiology, 2. Clinical grade, 3. Histological grade (evidences of inflammation and necrosis), and 4. The stage (extent of fibrosis and reflects the level of progression of the disease).

(D) Clinical features—

1. Fatigue, malaise, anorexia, fever.
2. Failure to recovery from acute hepatitis.
3. Jaundice (persistent or intermittent).
4. Hepatomegaly with or without splenomegaly.
5. Signs of hepato-cellular failure.

(E) Diagnosis—

1. High bilirubin level with very high aminotransferase levels; prolonged prothrombin time.
2. Antinuclear factor (ANF), anti-smooth muscle antibody may be present.
3. Hypoalbuminaemia with hyperglobulinaemia (autoimmune variety).
4. *Liver biopsy—diagnostic (described above in 'Types').*

Association of lymphadenopathy with jaundice :

Think of :

- | | |
|--|---------------------------------|
| 1. Lymphoma. | 4. Infectious mononucleosis. |
| 2. Leukaemia (ALL). | 5. Disseminated tuberculosis. |
| 3. Viral hepatitis (cervical and rarely epitrochlear). | 6. Disseminated carcinomatosis. |

How do you like to investigate a case of viral hepatitis ?

1. Blood for TC, DC and ESR—Leucopenia, lymphopenia and neutropenia. Blood picture becomes normal as jaundice appears. Atypical lymphocytes (virucytes) may be seen. ESR is high in pre-icteric phase and returns to normal with jaundice.
2. Liver function tests—
 - a) Serum bilirubin—High (in most instances the total bilirubin is equally divided between the conjugated and unconjugated fraction, or conjugated fraction is increased).
 - b) SGOT (AST) and SGPT (ALT)—Increased (normal value is 10-40 IU/L).
 - c) Prothrombin time—If increased, indicates extensive hepato-cellular necrosis and a worse prognosis. Prothrombin time is the best prognostic indicator of hepato-cellular failure.
 - d) Alkaline phosphatase—Slight increase (moderately increased in cholestasis; normal value is 35-130 IU/L).
 - e) Serum proteins—Mild elevation of gamma-globulin is common in acute viral hepatitis.
3. Urine and stool examination—
 - a) Urobilinogen in urine appears in pre-icteric phase, disappears as jaundice is evident and reappears with recovery.
 - b) Bilirubin appears in urine before clinical jaundice develops.
 - c) Rarely proteinuria may be present.
 - d) Stool—May reveal steatorrhoea.
4. Serological diagnosis—
 - a) HB_sAg (Australia antigen)—Positive in Type B (appears 6 weeks after infection and usually disappears by 3 months).
 - b) Anti-HAV of IgM class — Positive in Type A variety.
 - c) Anti-HCV (+ve after 6 weeks of infection; detected by third generation ELISA) and HCV RNA—Positive in Type C variety.
 - d) Anti-HDV and anti-HEV antibody of IgM type are present in hepatitis D and E respectively.
 - e) Serology for cytomegalovirus or Epstein-Barr virus.
5. Blood glucose level may be low (vomiting, i intake, poor hepatic glycogen).

6. Imaging—Ultrasonography (commonly used to differentiate other causes of jaundice) or CT scan of liver (USG is superior to CT scan).
7. Liver biopsy—Usually it is not required to diagnose acute viral hepatitis. It is important in differentiating cirrhosis, chronic hepatitis, drug-induced hepatitis from acute viral hepatitis.
8. Work-up to differentiate from cirrhosis of liver, alcoholic hepatitis (AST : ALT > 2 and high gamma-glutamyl transpeptidase), autoimmune chronic active hepatitis (enzymes >10 times and gamma globulin > 2 times of normal), hepatic amoebiasis (positive indirect haemagglutination test, chest X-ray) should be done.

N.B. : One should always search for K-F ring in the eyes in all cases of recurrent jaundice in young individuals (to exclude Wilson's disease producing cirrhosis of liver).

Common clinical differences between type A and type B infection :

Characteristics of type A infection are :

1. Fever may be high; > 102° F.
2. Onset Is very acute.
3. Cholestasis is more common.
4. Fulminant hepatitis is less common.
5. Less severity of illness with most patients making a complete recovery.
6. Post-hepatitis syndrome is more common.
7. No chronicity, no carrier state.
8. Simultaneous infection of other family members or persons in the same locality.
9. Serum sickness-like syndrome does not occur (very characteristic of type B).
10. Usually there is no past H/O blood transfusion, needle prick etc.

How to recognise that recovery has started in acute viral hepatitis ?

1. Appetite returns.
2. Stool becomes normal coloured (from white or clay colour).
3. Urine and eye become less yellowish.
4. Bilirubin and enzymes level return towards normal.

Who is a 'carrier' of type B infection ?

Persistence of HB_sAg for more than 6 months is known as 'carrier'.

How the ongoing infectivity and / or chronicity is diagnosed in type B infection ?

- a) Ongoing infectivity (viral replication) is diagnosed by the presence of HB_eAg and positive serum HBV DNA.
- b) Chronicity is diagnosed by the presence of positive serum IgG anti-HBc.

Discuss the post-exposure prophylaxis :

- (A) Hepatitis A—It is often necessary for household and institutional 'intimate' contacts. Immune globulin is administered in a dose of 0.02 ml/kg of body weight by I.M route. Hepatitis A vaccine (Havrix) can be given (0.5 ml), I.M, in 2 doses at 0, 6-12 months in 2-18 years age group; over 18 years, it is given (1 ml) by 2 injections in the same schedule of 0, 6-12 months.
 - (B) Hepatitis B—
 - (i) HBIG—0.5 ml (neonates) or 0.06 ml/kg (adults) as single dose immediately by I.M route, plus
 - (ii) Hepatitis B vaccine (Engerix-B, hepavax)—3 doses (each of 1 ml) are given by I.M route (always in deltoids) at 0, 1 and 6 months; accelerated schedule of 0, 1, 2, and 12 months are also approved (preferred). Booster dose is not considered routinely (protection appears to be excellent in normal schedule) except in immunocompromised and haemodialysis patients.
 - (C) Hepatitis C or E—No available active or passive protection. Development of vaccine is in progress.
- N.B. : Pregnancy is not a contraindication to vaccination against hepatitis B. Vaccination against hepatitis B will give protection from delta virus infection too. HBIG may be repeated at 4 weeks.

Indications for hepatitis B vaccination :

1. Surgical staffs, dental staffs and medical students.
2. Hospital and laboratory staffs in direct contact with blood.
3. Patients and staffs in haematology (haemophilia), oncology, and haemodialysis departments.
4. Mental subnormality (e.g., Down's syndrome).

5. I.V drug abusers.
 6. Homosexually and bisexually active men; prostitutes; HIV infection or AIDS.
 7. Babies born to HB_sAg + ve mother.
 8. Accidental exposure to HB_sAg + ve blood.
 9. Close family and sexual contacts of HB_sAg + ve carriers.
- * 1 to 6 require pre-exposure prophylaxis (0, 1, 6-month's schedule), and 7 to 9 require post-exposure prophylaxis (accelerated). In post-exposure prophylaxis, HBIG should be given along with the vaccine.

Who discovered Australia antigen ?

In the year 1965, Blumberg discovered the hepatitis B surface antigen (HB_sAg) and was awarded Nobel Prize for his discovery in 1977.

Diagnosis of hepatitis C virus infection :

1. Read the clinical characteristics mentioned earlier.
2. Serological test for HCV antibodies (anti-HCV) is usually positive after 6 weeks of infection.
3. Presence of HCV-RNA is detected by polymerase chain reaction (PCR). 1 -2 weeks after infection.

How will you manage your case ?

There is no specific treatment for acute viral hepatitis.

1. Bed rest with bathroom privileges till jaundice persists. Restrict physical activity.
 2. Diet—Low-fat, high carbohydrate, high-calorie diet (a diet containing 2000-3000 Kcal should be given). The food must be palatable. Plenty of fluids to drink.
 3. Avoid close physical contacts in acute stage. Avoid alcohol. sharing comb/razors. Regular surveillance on clinical parameters as well as liver function tests is done. Reassurance, as recovery is the rule in majority.
 4. Symptomatic treatment—
 - a) Antiemetics—Domperidone, metoclopramide or ondansetron may be used.
 - b) Sedatives—Should not be used. If the patient is very restless, oxazepam may be given.
 - c) Vitamins—Parenteral injection of vitamin K, 10 mg by I.M route may be given for consecutive 3 days if prothrombin time is high. Other vitamin supplements are not necessary.
 - d) Severe vomiting with fluid loss—10% dextrose drip may be given.
 - e) Cholestyramine or UDCA (ursodeoxycholic acid) is advocated for intense pruritus from cholestasis.
 5. Treatment of hepato-cellular failure (e.g., in fulminant hepatitis) may be done accordingly.
- * Steroids should not be used in acute viral hepatitis. Keep the patient on minimal drugs.

Patient of viral hepatitis becomes drowsy—significance :

Patient is going towards hepatic pre-coma (encephalopathy).

Case 15

Cirrhosis of Liver

What is your diagnosis ?

This is a case of chronic liver disease probably macronodular cirrhosis of liver with features of portal hypertension and hepato-cellular failure following type B hepatitis, and the disease is progressing.

- * Chronic liver disease is defined as liver diseases persisting for more than 6 months.

Why do you say so ?

This is a case of cirrhosis of liver because :

(A) From history :

- a) Fatigue, weight loss, flatulent dyspepsia, anorexia and low grade fever for last 6 months.
- b) Haematemesis, melaena, swelling of legs and progressive distension of abdomen for last 10 days,
- [c] Drowsiness, inversion of sleep rhythm for last 2 days (say, if present)].
- d) Past H/O hepatitis B infection 12 years back.

(B) Physical examination :

a) General survey—

1. Malnutrition.
2. Anaemia—Moderate.
3. Jaundice—Mild.
4. Hepatic facies.
5. Oedema feet.
6. Obvious swelling of abdomen.
7. K-F ring in the eyes—Absent.
8. Flapping tremor—absent.

Systemic examination —

1. Abdomen—

- (1) Venous prominences over abdomen, specially in the epigastrium with flow of blood away from the umbilicus.
- (ii) Liver is 1 cm palpable, firm, non-tender with sharp and irregular margin; chiefly the left lobe is palpable (liver may not be palpable, specially in late stages, when it shrinks in size due to progressive hepatocyte destruction and fibrosis).
- (iii) Moderate, non-tender splenomegaly.
- (iv) Ascites—Confirmed by shifting dullness.

2. Skin changes—

- (i) Spider naevi.
 - (ii) Palmar erythema.
 - (iii) Diffuse pigmentation of skin.
 - (iv) White nails or leuconychia (due to hypoalbuminaemia).
 - (v) Scanty axillary and pubic hair.
3. Gynaecomastia and bilateral testicular atrophy (soft and small testes), [or atrophy of breast (females)].
 4. Neurological changes—Nothing abnormal at present (say, if present).

Summary : This is a case of cirrhosis of liver because of—

- Long history
- H/O jaundice, haematemesis, melaena and oedema
- Poor nutritional status, and
- Features of portal hypertension and hepato-cellular failure.

What is your case ?

Build up the summary.

Importance of past history :

1. Jaundice (viral hepatitis).
2. Drugs (rifampicin, INH, anabolic steroids, OC pills; NSAID for melaena) or any herbal remedies taken.
3. Blood transfusion or transfusion of any blood products (viral hepatitis B or C; recent tattooing or acupuncture; drug abuse).
4. Alcohol consumption.
5. Tuberculosis (ascites due to tuberculous peritonitis).
6. Haematemesis or melaena (peptic ulcer, ruptured oesophageal varix, gastric malignancy).
7. Fever (tuberculosis, hepato-cellular failure etc).
8. Haematochezia (lower G. I. malignancy, haemorrhoids).

Queries in personal and family history :

Vide page 5 with special reference to alcohol intake. Dose of alcohol consumption, type of drink, daily/weekly pattern should be mentioned with duration. The patient takes mixed Indian diet Family history may be positive in Wilson's disease.

Importance of family history in alimentary system :

- Similar illness in the family—acute diarrhoea, food poisoning and viral hepatitis.

- Genetic hepatic disorders—Wilson's disease, haemochromatosis and α_1 -antitrypsin deficiency.
- Familial polyposis coli, inflammatory bowel disease and carcinoma of colon.

Features of portal hypertension :

1. Evidences of porto-systemic anastomosis (collateral circulation) at different sites :
 - a) Lower end of oesophagus—Rupture of 'oesophageal varix' produces haematemesis and melaena.
 - b) Around the umbilicus—Caput medusae (classical caput medusae is rare); and also in the anterior abdominal wall, specially in the epigastrium.
 - c) Rectum and anal canal—Haemorrhoids.
2. Splenomegaly (single most bedside cardinal finding), and
- 3 Ascites.

* It is often said that fetor hepaticus is a feature of portal hypertension (due to porto-systemic shunting of blood) but it should be remembered that underlying hepato-cellular failure must be present. *Portal hypertension is unlikely if splenic enlargement can not be demonstrated clinically or by radiographically.*

** Caput medusae—looks like Greek Goddess medusae's hair after minerva had turned it into snakes i.e., snakes radiated from head (caput) of Medusae.

Features of hepato-cellular failure :

1. General failure of health (loss of flesh).
2. Jaundice.
3. Skin changes (read the section on 'Palmar erythema' too)
 - a) Spider naevi.
 - b) Palmar erythema (liver palm).
 - c) Diffuse pigmentation.
 - d) White nails (due to hypoalbuminaemia).
 - e) Clubbing (common in biliary cirrhosis).
 - f) Loss of axillary and pubic hair (due to hyperoestrogenaemia); alopecia.
 - g) 'Paper money' skin (usually on upper arms).
 - h) White spots (arm and buttock).
4. Endocrine changes—
 - a) Gynaecomastia (breast atrophy in females)
 - b) Testicular atrophy (bilateral).
 - c) Menstrual irregularities in females.
5. Bleeding manifestations—Petechiae, ecchymosis (chiefly due to hypoprothrombinaemia and sometimes due to low platelet count).
- 6 Fever (related to endotoxaemia with production of cytokines).
7. Fetor hepaticus (sweetish-faecal smell of the breath and urine due to methyl mercaptan derived from methionine; the smell resembles that of dead mouse).
- 8 Hepatic encephalopathy (disturbed consciousness with inversion of sleep rhythm, personality changes intellectual deterioration, psychiatric abnormalities with asthenia)—personality changes e.g., irritability and loss of family concern, and constructional apraxia i.e., inability to reproduce simple diagrams with blocks are common.
9. Ascites and bipedal oedema.
10. Circulatory changes—
 - (A) Hyperkinetic circulation—
 - a) Capillary pulsation.
 - b) Bounding pulse (high pulse pressure with low diastolic BP).
 - c) Tachycardia.
 - d) Hyperdynamic apex.
 - e) Ejection systolic murmur at the apex.
 - (B) Cyanosis and clubbing—Due to development of pulmonary arteriovenous shunts.

* To diagnose a case of cirrhosis of liver, one should search for features of **both** portal hypertension and hepato-cellular failure. Pedal oedema (pitting) develops as a result of hypoproteinaemia, and additional factor is functional IVC obstruction due to ascites.

** _A female distribution of body hair may be seen in male cirrhotics (e.g., less frequent shaving).

*** Features accountable for both hepato-cellular failure and portal hypertension ; ascites and fetor hepaticus (according to some hepatologists).

What ascites indicates in cirrhosis of liver ?

It indicates hepato-cellular failure with portal hypertension, i.e., it is decompensated cirrhosis with portal hypertension.

Features of 'hyperoestrogenaemia' in hepato-cellular failure :

Diminished hepatic clearance of precursor androstenedione results in peripheral conversion to oestrogen, and manifests as—

1. Spider naevi.
2. Palmar erythema.
3. Gynaecomastia (breast atrophy in females).
4. Bilateral testicular atrophy and infertility.
5. Loss of beard (H/O less shaving), axillary and pubic hair.
6. Menstrual irregularities.
7. Loss of libido.

Describe 'hepatic fades' :

1. Shrunken eyes.
2. Hollowed temporal fossa.
3. Pinched-up nose with malar prominence.
4. Parched lips.
5. Muddy complexion of skin (blending of pallor, jaundice and melanosis).
6. Shallow and dry face.
7. Icteric tinge of conjunctiva.

Hepatic fades is characteristic of chronic liver disease e.g., cirrhosis of liver.

How to narrate a complete diagnosis in cirrhosis ?

(A) Aetiological Alcohol, type B hepatitis, Wilson's disease etc.

(B) Morphological—Micronodular, macronodular or mixed nodularity.

(C) Functional assessment—

1. Portal hypertension.
2. Hepato-cellular failure.
3. Evolution—Progressing, regressing or stationary.

Morphological classification of cirrhosis :

(B) Micronodular—Alcoholism, malnutrition, old age, anaemia, haemochromatosis.
Post-HBV, Post-HCV, Wilson's disease, a, -antit-

(C) MIXED—Features both micro- and macronodular cirrhosis e.g., biliary cirrhosis.

** Micronodular = Laennec's cirrhosis, and macronodular cirrhosis = postnecrotic or viral.

When regeneration is slow, micronodules are developed.

Special stigmata of alcoholic cirrhosis :

1. Bilateral enlarged parotids.
2. Gynaecomastia.
3. Testicular atrophy with loss of body hair.
4. Wasting of muscle mass, and
5. Dupuytren's contracture.

* Parotid cysts are seen in suppurative parotitis, mumps, cirrhosis of liver, Sjogren's syndrome, lymphomatous deposits, post-operative patients, calculi, drug-induced (iodides, guanethidine), parotid neoplasms or amyloidosis.

'Cirrhogenic' or danger dose of alcohol :

consumption is > 80 g/day. Most alcoholic cirrhotics consume approximately 80 g/day. A steady daily intake is more dangerous than intermittent drinking. Chance of alcohol-related liver damage is less if the intake is below 40 g/day.

Alcohol equivalents :

Whisky	30 ml	=	10 g.
Country liquor	45 ml	=	10 g.
Wine	100 ml	=	10 g.
Beer	250 ml	=	10 g.

Aetiology of cirrhosis of liver :

1. Alcoholic.
2. Biliary (primary or secondary).
3. Cryptogenic (when no aetiology can be determined).
4. Postnecrotic or postviral (HBV, HCV, HDV).
5. Cardiac or congestive.
6. Metabolic—Wilson's disease, haemochromatosis, type IV glycogenosis, α -antitrypsin deficiency, galactosaemia, cystic fibrosis.
7. Drugs—Methotrexate, amiodarone, methyldopa, halothane.
8. Miscellaneous—
 - (i) Indian childhood cirrhosis.
 - (ii) Budd-Chiari syndrome.
 - (iii) Intestinal by-pass surgery.
 - (iv) Autoimmune hepatitis (lupoid).
 - (v) Non-alcoholic steatohepatitis (NASH).
 - (vi) Primary sclerosing cholangitis.

Compensated and decompensated cirrhosis :

Chronic liver disease usually passes through a long period of minimum non-specific symptoms like fatigue, flatulent dyspepsia, anorexia—known as 'COMPENSATED' cirrhosis; and appearance of ascites, jaundice, encephalopathy, G.I. bleeding and pre-coma are known as 'DECOMPENSATED' cirrhosis.

Examination of eye in cirrhosis :

1. Icteric tinge (jaundice).
2. Anaemia.
3. Bitot's spot.
4. K-F ring.
5. Subconjunctival haemorrhage (due to hypoprothrombinaemia).

* Always search for K-F ring in a patient with cirrhosis of liver, specially in younger cirrhotics.

Sites for varices :

- | | |
|---|----------------------|
| 1. Lower end of oesophagus (commonest). | 3. Colo-rectal. |
| 2. Gastric varices. | 4. Rarely, duodenal. |

Describe the 'liver' in cirrhosis :

The liver may not be palpable in the presence of ascites. Usually the liver is,

1. Palpable, 1 cm below the right costal margin at right MCL (often the left lobe is better palpable in the epigastrium) and moving with respiration. The liver is enlarged in early cirrhosis and may be shrunken in advanced disease.
2. **Firm in consistency.**
3. **Sharp and irregular margin.**
4. Surface is finely irregular (portal or Laennec's cirrhosis) or coarsely irregular (postnecrotic).
5. Non-tender.
6. Upper border of liver dullness may be lowered (in 6th or 7th ICS at right MCL).
7. No bruit.
8. No rub.
9. No pulsation.

* There is **moderate splenomegaly in cirrhosis.**

Aetiology of portal hypertension :**(A) Pre-sinusoidal :**

- a) **EXTRAHEPATIC (portal vein thrombosis or extrahepatic obstruction or EHO)—**
 1. Umbilical sepsis in neonates.
 2. Portal pyaemia.
 3. Exchange transfusion in neonates.
 4. Oral contraceptives, pregnancy.
 5. Abdominal trauma.

6. Portal lymphadenopathy (lymphoma, metastasis).
 7. Hyperviscosity syndrome (myeloproliferative disorders).
 8. Malignancy of liver and pancreas.
 9. Protein C or S deficiency, antithrombin III deficiency, factor V Leiden.
 10. Secondary to cirrhosis of liver.
 11. Idiopathic,
- b) INTRAHEPATIC—
1. Schistosomiasis.
 2. Congenital hepatic fibrosis.
 3. Sarcoidosis.
 4. Lymphoma.
 5. Leukaemic infiltrations.
 6. Primary biliary cirrhosis.
 7. Arsenic, vinyl chloride and copper intoxication.
 8. Idiopathic portal hypertension.

(B) Post-sinusoidal :

- a) INTRAHEPATIC (better known as 'intrahepatic parenchymal')—
1. Cirrhosis of liver (commonest cause of portal hypertension).
 2. Acute alcoholic hepatitis.
 3. Hypervitaminosis A.
 4. Cytotoxic drugs.
 5. Veno-occlusive disease (by Jamaica bush tea containing toxic pyrrolizidine alkaloids).
 6. Partial nodular transformation of liver.
 7. Metastatic malignancy of liver.
- b) EXTRAHEPATIC—
1. Budd-Chiari syndrome (obstruction of hepatic veins at any site from efferent vein of the lobule to the entry of the IVC into right atrium)
 - (i) Pregnancy.
 - (ii) Oral contraceptives.
 - (iii) Abdominal trauma.
 - (iv) Malignancy of liver, kidney, adrenal, testis etc.
 - (v) Membranous obstruction of IVC (webs, diaphragms).
 - (vi) Hyperviscosity syndrome (myeloproliferative disorders).
 - (vii) Thrombophilic states (antiphospholipid syndrome, protein C or S deficiency).
 - (viii) Paroxysmal nocturnal haemoglobinuria.
 - (ix) Right atrial myxoma.
 - (x) Others—vasculitis (Behcet's disease), liver abscess, sickle cell disease.
 - (xi) Idiopathic.
 2. Constrictive pericarditis.
 3. Severe right heart failure.

* Actually, sinusoidal causes of portal hypertension are 1, 2, 6 and 7 of (B)-a).

** Two rare causes of portal hypertension are tropical splenomegaly syndrome and haematological disorders with massive splenomegaly (increases hepatic blood flow).

*** In Budd-Chiari syndrome, there are added features of inferior vena caval obstruction (distended veins over flanks and back, plus gross pedal oedema) over and above portal hypertension and hepatocellular failure.

Summary of aetiology of portal hypertension :

- Pre-hepatic—EHO i.e., portal vein thrombosis and splenic vein thrombosis
- Hepatic—
 - a) Pre-sinusoidal—non-cirrhotic portal fibrosis
 - b) Sinusoidal—cirrhosis of liver
 - c) Post-sinusoidal—veno-occlusive disease
- Post-hepatic—Budd-Chiari syndrome, constrictive pericarditis



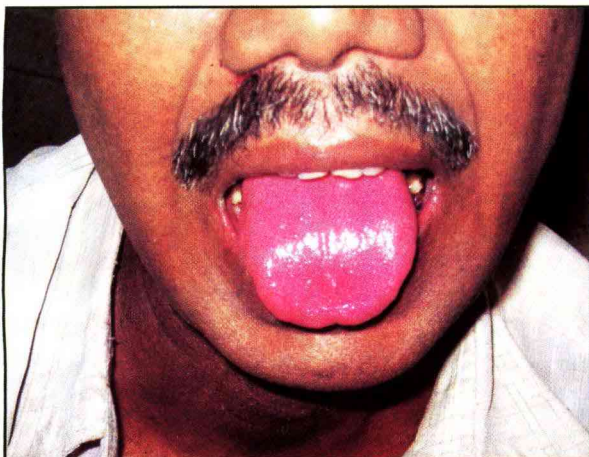
Ovoid shallow painful **aphthous ulcer** (canker sore) in tongue with a white centre and inflammatory halo



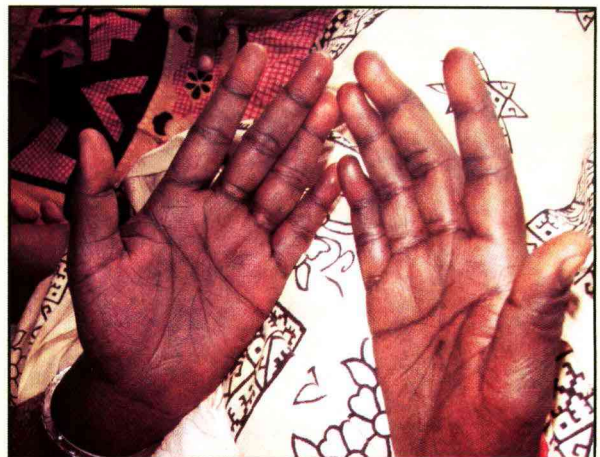
Ichthyosis (fish-scale skin) in extremities



← Flaccid and wasted **tongue** with **fasciculation** in bulbar palsy from chronic motor neurone disease



Magenta-coloured tongue (pink tongue with glossitis) with mild angular stomatitis in riboflavin deficiency



Black palm with **blackish palmar creases** in Addison's disease



Dermatomyositis with heliotrope (lilac-coloured) rash in eyelids, periorbital oedema, and purple pigmentation in forehead, nose, chest and upper arm



Systemic lupus erythematosus with classical 'butterfly' rash in the face



Scleroderma showing mask-like facies, pinched up nose, taut and shiny skin over the face. Pigmentation and depigmentation are also noted



A **leprosy** patient showing thickened skin of face and forehead, especially the infiltrated earlobes



Parotid enlargement by leukaemic deposits in right parotid gland in acute lymphoblastic leukaemia; pallor of the face can not be overlooked

What is non-cirrhotic portal fibrosis (NCPF) ?

NCPF is an intrahepatic pre-sinusoidal cause of portal hypertension. The disease is characterised by moderate to massive splenomegaly with anaemia, 'preserved hepato-cellular function' and a benign prognosis in majority though in the late stages, nodular transformation may occur. Though recurrent haematemesis is common and a presenting feature, hepatic encephalopathy is uncommon. Ascites is unusual in NCPF. Endoscopy reveals oesophageal varices, splenoportalvenography (SPV) shows massive dilatation of portal and splenic veins with presence of collaterals; liver biopsy demonstrates varying degree of portal fibrosis with maintained lobular architecture. The possible aetiologies are :

1. Idiopathic portal hypertension (previously known as Banti's syndrome),
2. Schistosomiasis,
3. Congenital hepatic fibrosis, and
4. Chronic arsenic ingestion (mainly through tube-well water; common in India and Bangladesh).

Features of extrahepatic obstruction (EHO) :

Patients presents at an earlier age (causes mentioned in page 123-124) with episodes of haematemesis and/or melaena, not accompanied by hepatic decompensation. Signs of stigmata of cirrhosis, jaundice, ascites, and hepatic coma from bleeding are uncommon. Liver is usually soft, if at all palpable There is massive splenomegaly with signs of portal hypertension. SPV demonstrates the collaterals and feeders vessels.

* Splenomegaly in different types of portal hypertension are :

- a) Cirrhosis—moderate, b) NCPF—upto midline, c) EHO—massive.

What is NASH or NAFLD ?

Diffuse accumulation of neutral fat (triglycerides) in hepatocytes may give rise to mild to moderate enlargement of liver, and is known as non-alcoholic steatohepatitis (NASH) or non-alcoholic fatty liver disease (NAFLD). 'Fatty liver' is divided into 2 types :

1. Macrovesicular (large fat droplets in hepatocytes)—alcohol, obesity, diabetes mellitus, starvation, drugs (steroid, oestrogen, amiodarone), total parenteral nutrition, jejunoileal by-pass, Indian childhood cirrhosis.
2. Microvesicular Reye's syndrome, acute fatty liver of pregnancy, drugs (tetracycline, valproate), toxins (yellow phosphorus).

Most patients are asymptomatic and may only complain of mild right hypochondriac discomfort. Hepatomegaly and mild elevation of aminotransferase are common. Fatty liver is reversible but if untreated there is chance of development of cirrhosis of liver (9-25%) in the long run. Insulin resistance is present in virtually all patients of NASH /NAFLD, and NAFLD is now considered as hepatic component of metabolic syndrome. Ursodeoxycholic acid (UDCA) and pioglitazone/rosiglitazone are used as pharmacotherapy of fatty liver. The patients are encouraged to lose weight.

Pain abdomen in cirrhosis :

1. Tuberculous peritonitis.
2. Spontaneous bacterial peritonitis (SBP) — infection of ascitic fluid without any obvious orimary source of infection.
3. Peptic ulcer (common in cirrhotics).
4. Chronic cholecystitis (1/3rd of cirrhotics have pigment gall stones).
5. Portal vein thrombosis.
6. Pancreatitis (common in alcoholic cirrhosis).
7. Cirrhosis turned into hepato-cellular carcinoma (hepatoma).
8. Zieve's syndrome (haemolysis, pain abdomen, hyperlipidaemia in an alcoholic patient).

Fever in cirrhosis :

- | | |
|---|---|
| 1. Tuberculous peritonitis. | 5. Associated chronic active hepatitis. |
| 2. Spontaneous bacterial peritonitis (SBP). | 6. Tuberculosis (pulmonary or intestinal). |
| 3. Hepato-cellular failure. | 7. Other causes of fever like malaria, enteric fever etc. |
| 4. Transformation into hepatoma. | |

Haematemesis and / or melaena in cirrhosis :

- | | |
|--|--------------------------------|
| 1. Ruptured oesophageal varices (commonest). | 5. Rupture of gastric varices. |
| 2. Peptic ulcer. | 6. Mallory-Weiss tear. |
| 3. Portal hypertensive gastropathy. | 7. Bleeding tendency. |
| 4. Gastric erosion (NSAID-induced). | 8. Ulcer due to sclerotherapy. |

* In approximately 50% cases, the source of upper G. I. bleeding in cirrhosis is from peptic ulcer or portal hypertensive gastropathy.

Definition of cirrhosis of liver :

Cirrhosis is a pathological term. It is the chronic and diffuse involvement of liver parenchyma of varied aetiology, and is clinically characterised by features of portal hypertension and hepato-cellular failure in varying combination, and pathologically by irreversible NECROSIS, extensive FIBROSIS as well as REGENERATIVE NODULE FORMATION in such a way that the normal architecture of liver is totally lost.

Importance of examination of other systems in cirrhosis :

- (A) RESPIRATORY SYSTEM—
 - a) Evidence of hydrothorax (commonly right-sided).
 - b) Evidence of pulmonary tuberculosis (common in cirrhosis).
- (B) CVS—Evidence of circulatory changes, already mentioned in 'features of hepato-cellular failure'.
- (C) NERVOUS SYSTEM—Neuropsychiatric manifestations of hepato-cellular failure (hepatic encephalopathy) are described below (see page 128).
- (D) GENITOURINARY SYSTEM—
 - a) Facial puffiness due to renal failure (hepato-renal syndrome) with oliguria.
 - b) Testicular atrophy and loss of pubic hair.

Causes of haematemesis :

(A) **COMMON CAUSES :**

- | | |
|--|----------------------------|
| 1. Chronic duodenal ulcer (commonest cause). | 6. Mallory-Weiss syndrome. |
| 2. Erosive gastritis. | 7. Peptic oesophagitis. |
| 3. Variceal bleeding (ruptured oesophageal varices). | 8. Duodenal erosions. |
| 4. Chronic gastric ulcer. | 9. Oesophageal carcinoma. |
| 5. Carcinoma of the stomach. | 10. Hiatal hernia. |

(B) **LESS COMMON CAUSES :**

- | | |
|--|---|
| 1. Leiomyoma and leiomyosarcoma of the stomach. | 11. Arteriovenous malformations or Dieulafoy's lesion. |
| 2. Lymphoma of the stomach. | 12. Corrosive ingestion (e.g., strong acid or strong alkali). |
| 3. Duodenal diverticula. | 13. 'Spurious' haematemesis (e.g., swallowed nose bleed). |
| 4. Blood dyscrasias (leukaemia, thrombocytopenia). | 14. Aorto-duodenal fistula; haemobilia. |
| 5. Anticoagulant therapy. | 15. Watermelon stomach (gastric antral vascular ectasia). |
| 6. Rendu-Weber-Osler disease. | 16. Idiopathic. |
| 7. Pseudoxanthoma elasticum. | |
| 8. Uraemia. | |
| 9. Amyloidosis. | |
| 10. Vasculitis. | |

* All the above mentioned causes may produce **melaena**.

** Dieulafoy's lesion—easily overlooked, small, superficial mucosal defect of stomach that involves an end artery which remains as large as its feeding artery -> pressure erosion of the epithelium by the vessel -> ultimately the vessel ruptures.

Describe the characteristic features of melaena :

Melaena is 'altered blood in stool' with the following characteristics :

1. Black tarry stool (due to production of acid haematin); sticky too.
2. Offensive (acid haematin is altered by bacteria).
3. Semisolid in consistency.
4. Red-coloured fluid comes out from the stool after addition of water in it.
5. Usually associated with vertigo, dizziness or syncopal attack during defecation.

Approximately 60 ml of blood is required to produce a single black stool and blood should remain at least 8 hours within the gut lumen to produce melaena. Haematemesis and melaena usually denote bleeding from the proximal gut (**proximal to the ligament of Treitz**) but bleeding from the lower gut may rarely produce melaena provided the gastrointestinal transit time is sufficiently prolonged. Occult blood test in stool may remain positive for seven days even after the colour of stool becomes normal. Majority of patients with haematemesis usually have melaena but less than 50% patients with melaena have haematemesis. Rarely, 'swallowed blood' may give rise to melaena (spurious) e.g., epistaxis.

What is haematochezia ?

It is the passage of bright red blood per rectum mixed with or without stool. It signifies bleeding from a source which is usually **distal to the ligament of Treitz**. The common examples of lower G. I. tract bleeding are haemorrhoids, anal fissure and fistula, trauma, proctitis, ischaemic colitis, ulcerative colitis, diverticulitis, polyp, malignancy of colon and rectum, angiodysplasia of colon, arteriovenous malformations, pseudomembranous colitis and 'gay bowel syndrome' in male homosexuals. Massive upper G.I. bleed may give rise to bright or dark red 'maroon' coloured stool, if there is hurried peristalsis or the transit time is reduced.

Causes of black stool :

1. Melaena.
 2. Ingestion of iron (as a haematinic)—Usually associated with hard stool.
 3. Ingestion of bismuth (in the treatment of chronic duodenal ulcer).
 4. Intake of licorice, charcoal (used in treatment of poisoning) or black berries.
- No. 2, 3 and 4 stools are non-sticky, and often known as pseudomelaena.

Blood in stool—what are the different forms ?

- 1) Frank blood or haematochezia.
- 2) Altered blood or melaena.
- 3) Invisible blood or occult blood (detected chemically).

Intake of NSAID, hookworm infestation and colo-rectal cancer are common causes of occult blood in stool.

- Obscure G.I. bleeding—where the cause of G.I. bleeding (haematemesis, melaena and haematochezia) is not found after proper and extensive investigations.

Prerequisites for occult blood test (guaiac test) in stool :

Bleeding from G.I. tract may be intermittent and thus the test should be performed for several (usually three) consecutive days.

1. 3 days high-fibre and meat-free diet (to avoid false positive result) is advocated.
2. Patient should not have been taking vitamin C (may result in false negative test).
3. Intake of NSAID and iron should be stopped. The patient may avoid tooth-brush for 3 days.

Indications of blood transfusion in haematemesis and / or melaena :

Transfusion is required if,

1. Patient is in persistent shock.
2. There is H/O syncope.
3. Pulse rate is > 100/minute.
4. Systolic BP is < 100 mm of Hg.
5. Haemoglobin concentration is < 10 g/dl.

Definition of portal hypertension :

Portal vein is a 5.5-8 cm long vein formed by the union of superior mesenteric vein and splenic vein, behind the head of the pancreas at about the level of L₂ vertebra. Approximately, 2/3rd of hepatic blood flow and 1 / 2 of the O₂ supply to the liver are provided by the portal vein.

Normal pressure in the portal vein is 7-10 mm of Hg or 10-15 cm of saline. Portal hypertension is defined as sustained elevation of portal pressure > 30 cm of saline (measured at surgery) or a direct percutaneous transhepatic portal vein pressure >12 mm of Hg.

How the oesophageal varices, caput medusae and haemorrhoids are formed in cirrhosis ?**• Oesophageal varices :**

- (A) Portal venous system—Left gastric vein, short gastric vein and posterior gastric veins communicate with,
- (B) Caval venous system—Intercostal veins, diaphragmo-oesophageal vein and azygos minor vein.

• Caput medusae (around umbilicus) :

- (A) Portal venous system—Paraumbilical veins communicate with,
- (B) Caval venous system—Superficial veins of the anterior abdominal wall.

• Internal haemorrhoids :

- (A) Portal venous system—Superior rectal vein communicates with,
- (B) Caval venous system—Middle and inferior rectal veins.

'Predictors' of bleeding from varices and variceal 'grading' :

(A) Best predictors of bleeding from varices are :

1. Size of the varix (larger the size, the more likely it is to bleed),
- * 2. Presence of red signs (red colour correlates with the flow of blood through the varix), and
3. Hepato-cellular function (hepatic failure promotes bleeding).

(B) Grading of varices (endoscopic) :

1. Grade 1 — the varices can be easily depressed by the endoscope.
2. Grade 2 — the varices can not be depressed by the endoscope.
3. Grade 3 — the varices are confluent around the circumference of the oesophagus.

* 'Cherry-red spots' viewed on endoscopy suggest imminent rupture of varices.

What is Child-Pugh classification in cirrhosis ?

In cirrhosis, Child's grade is used to assess the hepato-cellular function. The determining 5 factors are : serum bilirubin, serum albumin, ascites, hepatic encephalopathy and prothrombin time. Every patient is assigned a class (A, B and C). Class A is 'prognostically' better and class C is the worst. Child-Pugh scoring system (scores 5-15 encompassing class A, B, C) is used to assess prognosis and is a reliable predictor of survival.

Anaemia in cirrhosis :

The possible explanations are—

1. Haematemesis and/or melaena.
2. Anorexia producing malnutrition.
3. Malabsorption.
4. Hypersplenism.
5. Haemolysis in alcoholic cirrhosis (along with hyperlipidaemia, it is known as Zieve's syndrome).

Jaundice in cirrhosis :

1. Impaired bilirubin metabolism (commonest).
2. Intrahepatic cholestasis.
3. Haemolysis (rare).
4. Viral hepatitis from blood transfusion (given to combat haematemesis or melaena).

How to suspect that cirrhosis has transformed into hepatoma ?

1. Rapid deterioration of general health.
2. Pain in right upper abdomen appears; fever may be there.
3. Rapidly developing huge ascites, not responding to conventional therapy.
4. Progressive hepatomegaly with hard and nodular liver.
5. Bruit may be audible (due to T vascularity); rub is not uncommon.
6. Increase in serum alkaline phosphatase and a-fetoprotein.

Features of hepatic encephalopathy :

(A) STAGE OF PRE-COMA :

a) Symptoms—

- (i) Alteration in behaviour (may be restless or violent initially).
- (ii) Impairment of memory and other intellectual functions.
- (iii) Inability to concentrate, apathy, slowness and brevity of response.
- (iv) Inversion of sleep rhythm (daytime sleep with arousal at night) and hypersomnia (one of the early changes).
- (v) Change of personality.
- (vi) Drowsiness, disorientation and confusion.
- (vii) Slurred and monotonous speech.
- (viii) Rarely, convulsions.

b) Signs—

- (i) Higher function—Disorientation in time, place and person; dysarthria.
- (ii) Flapping tremor (asterixis); handwriting—Micrographia.
- (iii) Constructional apraxia (elicited by positive Reitan trail-making test).
- (iv) Rigidity.
- (v) Ankle clonus—May be present.

(vi) Plantar response—Flexor.

(vii) Gait—Ataxic.

(B) *STAGE OF HEPATIC COMA :*

- a) Patient is in deep coma.
- b) Tone of muscles—Flaccid.
- c) Jerks or deep reflexes—Absent.
- d) Plantar response—Extensor.
- e) Flapping tremor — Absent.

* EEG (electroencephalogram) shows symmetric high voltage triphasic waves changing down to the delta ranges (slow activity).

** 'Minimal' hepatic encephalopathy : few patients are clinically normal but show significant impairment in their psychometric performance and electrophysiological variables including the EEG.

Precipitating factors for hepatic encephalopathy in cirrhosis :

1. Gastrointestinal bleeding (from ruptured varices or peptic ulcer).
2. Excess protein in diet.
3. Overzealous use of diuretics (produces hypokalaemia alkalosis; NH_3 can not be converted into NH_4^+ ; so, hypokalaemia is very dangerous in cirrhotic patient).
4. Infection (sepsis).
5. Surgery (even in an operation on a small abscess).
6. Paracentesis abdominis (volume > 3-5 litre).
7. Sedatives, hypnotics.
8. Acute alcoholic bout.
9. Porto-systemic shunts made surgically or formed spontaneously (large shunts).
10. Constipation; vomiting and diarrhoea (produces dyselectrolytaemia).
11. Uraemia.

* Anaemia, hypoxia, hypoglycaemia and hypotension are also important precipitating factors.

Aetiopathogenesis of hepatic encephalopathy :

The different hypothesis are :

1. Increase ammonia level in blood (most important)— NH_3 combines with α -ketoglutaric acid in the Krebs's cycle and forms glutamic acid. So there is substrate deficiency in Krebs's cycle. NH_3 is the most readily identified toxin though it is not solely responsible for altered mental status.
2. Increase in short-chain fatty acids.
3. Rise in false neurotransmitters like octopamine.
4. Rise in methionine level.
5. Increased sensitivity of CNS neurones to gamma-aminobutyric acid (GABA), an inhibitory true neurotransmitter in the CNS.
6. Increased conversion of tryptophan to inhibitory neurotransmitter serotonin.
7. Ratio of aromatic amino acids (phenylalanine, tyrosine and tryptophan) to branched-chain amino acids (leucine, isoleucine and valine) is raised.
8. Excessive manganese deposition in the basal ganglia.
9. Increased circulating levels of endogenous benzodiazepines.

* Two general factors responsible for hepatic encephalopathy are liver failure and shunting of portal blood directly to the systemic circulation by-passing the liver.

** NH_3 also depresses cerebral blood flow and glucose metabolism.

Define fulminant hepatic failure :

It is a clinical syndrome resulting from massive necrosis of liver cells with sudden and severe impairment of hepatic function (it is virtually a medical hepatectomy). The definition usually includes a time frame of 12 weeks from the onset of jaundice, in patients without pre-existing liver disease.

1. Fulminant—hepatic failure develops within 2 weeks from the onset of jaundice.
2. Sub-fulminant—hepatic failure develops between 2-12 weeks of the appearance of jaundice.

Hepatic failure has been recently classified as,

- a) Acute (0-4 wks)
 - (i) Hyperacute (0-7 days),
 - (ii) Acute (8 days - 4 wks)
- b) Subacute (29 days - 12 wks)
- c) Chronic (> 24 wks)

Common causes of acute (fulminant) hepatic failure :

The causes vary throughout the world and they are—

- | | |
|--|------------------------------------|
| 1. Acute viral hepatitis (commonest). | 6. Shock and multi-organ failure |
| 2. Drug-induced (halothane, rifampicin-INH combination, paracetamol overdose). | 7. Wilson's disease. |
| 3. Mushroom or copper sulphate poisoning. | 8. Acute fatty liver of pregnancy. |
| 4. Weil's disease. | 9. Reye's syndrome. |
| 5. Acute Budd-Chiari syndrome. | 10. Autoimmune hepatitis. |

D/D you like to consider in your case :**(A) On aetiological basis :**

- Cryptogenic cirrhosis—This term is now used interchangeably with postnecrotic cirrhosis (the case described at the outset) but the terminology should be reserved for the patients where the aetiology is not known (10% of all cirrhotics). Clinical presentation in both the types are almost same. In many cases of cryptogenic cirrhosis, unrecognised NASH/NAFLD may play a role.
- Alcoholic cirrhosis—Male preponderance, H/O intake of alcohol in cirrhotic dose (commonly one pint or more of alcohol taken daily for 10 years), jaundice is less persistent, mild ascites, alcoholic stigmata present, H/O pancreatitis and peptic ulcer; spleen is less commonly palpable.
- Primary biliary cirrhosis—Commonly in middle-aged woman with persistent generalised pruritus, dark urine and pale stool, jaundice, clubbing, pigmentation, xanthelasma and xanthomas, gradual development of ascites, oedema and hepato-cellular failure.
- Wilson's disease—Young patient with hepatosplenomegaly and K-F ring in cornea; neurological manifestations like chorea, slurred speech, tremor are not uncommon. Positive family history may be obtained.
- Indian childhood cirrhosis—Age group is 6 months to 3 years, onset around first year of life with progressive distension of abdomen, irritability, low grade fever, firm liver with 'sharp leafy margin', splenomegaly with progressive hepato-cellular failure.
- α_1 -antitrypsin deficiency—Young age group, patient (PiZZ phenotype) usually suffers from respiratory distress due to emphysema or bronchiectasis along with chronic liver disease.
- Secondary biliary cirrhosis—H/O previous surgery in biliary tract with H/O ascending cholangitis. There is presence of pain in the right hypochondrium. This type of cirrhosis results from partial or total obstruction of the CBD for more than one year. Other clinical features are like that of primary biliary cirrhosis.

(B) D/D done by :

- Hepatomegaly.
- Splenomegaly.
- Ascites (never forget to make D/D with constrictive pericarditis).
- Causes of haematemesis and melaena.
- D/D of jaundice.

* Mention the D/D of your case according to the findings elicited and then apply your judgement. Cirrhosis of liver should be read along with the section on 'Ascites'.

What is cardiac cirrhosis ?

Prolonged passive venous congestion to liver from valvular heart disease, constrictive pericarditis or long-duration congestive cardiac failure may lead to cirrhotic changes in liver, and is known as cardiac cirrhosis. Clinical examination shows firm, non-tender and non-pulsatile hepatomegaly, features of right-sided heart failure, and stigmata of chronic encephalopathy with waxing and waning. This is a very rare cause of cirrhosis in clinical practice.

Can it be a case of carcinoma of liver ?

No. Characteristics of carcinoma of liver are :

- Cachexia, loss of weight, very rapid course.
- Large, tender, grossly nodular, hard liver with bruit or hepatic rub.
- Absence of splenomegaly.**
- Jaundice is of variable degree (secondary carcinoma of liver may be present with jaundice; hepatoma usually presents without jaundice).
- Ascites (often tender) with pedal oedema.
- Neck glands may be palpable.
- Usually there is no porto-systemic venous collaterals.

* Secondary carcinoma of liver is seen in the aged patients while hepatoma may affect persons between 40-50 years of age.

Nodular ***liver, jaundice, splenomegaly, ascites : can it be carcinoma of liver ?***

In carcinoma of liver there is absence of splenomegaly. Rarely, this case may be a primary hepatocellular carcinoma (hepatoma) superimposed on cirrhosis of liver. Presence of splenomegaly indicates the origin from cirrhosis of liver.

Portal hypertension with massive splenomegaly :

Portal hypertension is usually associated with moderate splenomegaly. Massive splenomegaly may be seen in,

1. Extrahepatic obstruction (i.e., presinusoidal-extrahepatic causes).
2. Cirrhosis with hypersplenism.
3. Non-cirrhotic portal fibrosis (NCPF).
4. Rarely in tropical splenomegaly syndrome.

Complications of cirrhosis :

1. Portal hypertension (variceal bleeding, congestive gastropathy, hypersplenism etc.).
2. Hepato-cellular failure and hepatic encephalopathy.
3. Ascites and spontaneous bacterial peritonitis (SBP).
4. Hepato-renal syndrome (functional renal failure with normal tubular functions develop due to circulatory or haemodynamic changes).
5. Acute and chronic pancreatitis.
6. Cholecystitis with pigment stone.
7. Peptic ulcer.
8. Septicaemia (gram-negative) and tuberculosis.
9. Portal vein thrombosis.
10. Hepato-cellular carcinoma (hepatoma) as a late complication.
11. Coagulopathy (factor deficiency, thrombocytopenia).
12. Hepato-pulmonary syndrome (HPS).
13. Porto-pulmonary hypertension.
14. Haematological complications (anaemia, haemolysis, thrombocytopenia).

* Hepato-pulmonary syndrome (HPS)—Cirrhotics may be hypoxaemic due to hepato-pulmonary syndrome (due to intrapulmonary shunting through direct arteriovenous communications -> may be a result of NO overproduction), porto-pulmonary hypertension (pulmonary hypertension as a result of vasoconstriction and obliteration of pulmonary arterial system —• leading to dyspnoea and fatigue), and hydrothorax. HPS is manifested by orthodeoxia, breathlessness, clubbing, cyanosis and spider naevi.

How do you suspect that spontaneous bacterial peritonitis (SBP) has developed?

SBP is suspected by appearance of fever, abdominal pain, increasing ascites not responding to diuretics, diminished bowel sounds and appearance of features of hepatic encephalopathy. It is an acute emergency and should be treated urgently.

Sudden worsening (decompensation) of stable cirrhosis—reasons behind :

1. Consider the precipitating factors for hepatic encephalopathy e.g., bleeding, hypokalaemia.
2. Development of spontaneous bacterial peritonitis or tuberculous peritonitis.
3. Transformation into hepatoma.
4. Formation of chylous ascites as a result of rupture of dilated abdominal lymphatics.
5. Portal vein thrombosis.

Possible causes of death in cirrhosis :

- | | |
|--|---------------------------------|
| 1. Hepatic encephalopathy. | 5. Coagulopathy. |
| 2. Cerebral oedema. | 6. Renal failure. |
| 3. Intercurrent infection (septicaemia). | 7. Hypoglycaemia, hypokalaemia. |
| 4. Respiratory failure. | 8. Acid-base imbalance. |

Renal failure in cirrhosis :

Renal failure in a patient with hepato-cellular failure may be due to various factors, e.g.,

1. Acute tubular necrosis precipitated by infection, haemorrhage or drugs (e.g., neomycin),
2. If associated with primary kidney disease.

3. Hepato-renal syndrome—When there is change in blood volume or shifting of fluid within the body compartment, this syndrome precipitates. The syndrome is due to increased pre-glomerular vascular resistance and thus the blood flow is diverted away from the renal cortex with resultant reduced GFR. The kidney are structurally normal but functionally abnormal.

Investigations you like to perform in your case :

1. Blood examination—TC, DC, ESR, Hb%.
 - a) Microcytic-hypochromic anaemia due to haematemesis and melaena.
 - b) Pancytopenia may be seen in hypersplenism.
 - c) ESR is increased in the presence of infection (like tuberculosis, SBP).
2. Stool for occult blood test—May be positive.
3. Rectal examination and proctoscopy for demonstration of internal haemorrhoids; slit-lamp examination of cornea for detection of K-F ring.
4. Chest X-ray—May show tuberculosis or hydrothorax.
5. Liver function tests—
 - a) *Total protein is diminished with low albumin and high globulin.* Albumin and globulin ratio is altered (normal albumin level is 3.5-5.5 g/dl and that of globulin is 2-3.5 g/dl). *Albumin* is due to impairment of hepatic protein synthesis whereas *T globulin* results from non-specific stimulation of reticulo-endothelial system.
 - b) Serum bilirubin—Normal or raised.
 - c) SGOT and SGPT (AST and ALT)—Mild elevation.
 - d) Alkaline phosphatase—Slightly increased.
 - e) Serum cholesterol—Low.
 - f) Prothrombin time—Increased and does not return to normal level with vitamin K therapy (it is a very important 'prognostic marker' of hepato-cellular damage. Normal prothrombin time is 12-16 seconds). Increased prothrombin time is a bad prognostic sign.
6. Serum autoantibodies—antinuclear antibody, anti-smooth muscle antibody and anti-mitochondrial antibody may be present in varying percentage in autoimmune hepatitis, primary biliary cirrhosis and cryptogenic cirrhosis.
7. Serum immunoglobulins are increased, e.g., IgG in autoimmune hepatitis, IgA in alcoholic cirrhosis and IgM in primary biliary cirrhosis.
8. Barium swallow X-ray of oesophagus demonstrates 'worm-eaten appearance' (multiple filling defects) at the lower part of oesophagus due to the presence of oesophageal varices. Sometimes, gastric varices may be seen by barium meal examination.
9. Endoscopy of upper G.I. tract—Shows oesophageal or gastric varices, peptic ulcer or congestive gastropathy (looks beefy-red: mosaic pattern of red and yellow mucosa with petechial haemorrhage). It is a very important and reliable primary investigation for portal hypertension.
10. Straight X-ray of abdomen—Usually not informative. It may show increased hepatic and splenic size or ground-glass opacity of ascites.
11. Examination of ascitic fluid—Transudate in nature if not complicated by tuberculosis or SBP. Serum-ascites albumin gradient (SAAG) is >1 g/dl (indicates underlying portal hypertension).
12. Ultrasonography (or CT scan) of liver, spleen, portal vein (size), any collaterals, presence of thrombosis (in portal vein, splenic vein, hepatic vein, IVC), and for any free fluid in the abdomen—in cirrhosis of liver, there is heterogenous echo-pattern of liver, splenic enlargement, presence of ascites and increase in diameter of portal vein (>13 mm). USG is superior to CT scan in cirrhosis.
13. **Liver biopsy—The gold standard test and confirms the diagnosis of cirrhosis of liver** (by Vim-Silverman, Menghini or Trucut' needle). Biopsy specimen can be specially stained for copper and iron.
14. Percutaneous intrasplenic pressure measurement—increased in portal hypertension.
15. Percutaneous transhepatic portal venography or splenoportalvenography (SPV)—For the demonstration of site of obstruction and nature of collaterals. The intrahepatic pattern in cirrhosis is known as 'tree in winter' appearance; 'cut off sign' is observed in NCPF.
16. Wedged hepatic venous pressure—Increased in cirrhosis of liver (never increased in NCPF).
17. Special investigations where facilities are available :
 - a) Visualisation of portal venous system by doppler ultrasound.

- b) Duplex doppler for measurement of portal blood flow,
- e) Portal venography by digital subtraction angiography.

18. Electroencephalogram (EEG)—Triphasic waves are seen in fulminant hepatic failure.

* Serum copper and ceruloplasmin estimation may be done for detection of Wilson's disease, serum iron for haemochromatosis, and serum α_1 -antitrypsin for aetiological diagnosis.

Management of cirrhosis of liver :

1. Rest—Absolute bed rest in decompensated stage (reduction of physical activity reduces metabolic demands of liver and increases the renal perfusion).
2. Diet—Salt-free high protein diet ($> 1\text{g/kg/day}$). Protein is restricted judiciously at the onset of hepatic encephalopathy.
3. Treatment of infection by proper antibiotics. SBP is treated by co-amoxycylav (amoxycillin 1g, clavulanic acid 200 mg, I.V, 8-12 hourly), or cefotaxime (1 g, I.V, 12 hourly) plus gentamycin (60 mg, I.V, 8 hourly), or piperacillin-tazobactam (4.5 g, I.V. 6 hourly) for 5-7 day.
4. Treatment of variceal haemorrhage by,
 - a) Blood transfusion.
 - b) Vasopressin, terlipressin, octreotide or infusion of somatostatin.
 - c) Sengstaken tube.
 - d) Endoscopic sclerotherapy of varices or band ligation.
 - e) Transjugular intrahepatic portosystemic stent shunting (TIPSS).
 - f) Lastly, oesophageal transection.
5. Treatment of hepatic encephalopathy by,
 - a) Protein-restricted diet. Vegetable protein is less ammoniagenic, better tolerated and more laxative.
 - b) Lactulose—10-30 ml three times daily (at least two to three semisolid stools per day). Lactitol may be used in lactulose-intolerant patients.
 - c) Neomycin—4g/day or metronidazole 200 mg, QDS, orally daily for 5-7 days. Rifaximin (1200 mg/day) is also effective.
 - d) Enemata : neutral phosphate, magnesium sulphate, mannitol; administer 8-12 hourly for 2-3 days.
 - e) L-ornithine L-aspartate (LOLA) given parenterally or orally promotes hepatic removal along with detoxification of NH_3 .
 - f) Maintenance of calorie, fluid and electrolytes.
6. Injection of vitamin K for coagulopathy—1 amp. of inj. vit. K, I.M daily for consecutive 3 days.
7. Judicious use of sedatives, if necessary—**safe sedatives** in liver disease are oxazepam, temazepam, heminevrin, phenobarbitone etc (morphine or paraldehyde are absolutely contraindicated).
8. Management of ascites by paracentesis, diuretics; control of oedema.
9. Control of aetiological factors :
 - a) Abstinence from alcohol intake.
 - b) Penicillamine in Wilson's disease.
 - c) Phlebotomy or desferrioxamine in haemochromatosis.
10. Management of fulminant hepatic failure in an 'intensive care unit' with experienced management team.

* Inj. ranitidine 50 mg, I.V, 6-8 hourly is given; dialysis is done for renal failure. Fresh frozen plasma is given if prothrombin time is > 1.5 times more than normal.

** Safest analgesic in cirrhosis is paracetamol. Neomycin is available as 350 mg tablet and avoided in the presence of renal failure.

*** Gross protein restriction as first-line treatment is not recommended now-a-days as it may lead to deteriorating nutritional state in already malnourished patients.

**** Prevention of recurrent variceal haemorrhage is done by sclerotherapy, banding, TIPSS, propranolol and porto-systemic shunt surgery.

How lactulose acts in hepatic encephalopathy?

Lactulose is a synthetic disaccharide. In the caecum, it is broken down into acetic acid and lactic acid, and thus the fecal pH drops. Its mechanism of action are ;

1. Produces acidic diarrhoea (acts as an enema or bowel wash).
2. As intestinal pH is acidic, NH_3 absorption is less (NH_3 is converted into NH_4^+).

3. Promotes incorporation of N_2 within the intestinal bacteria.
4. The drop in fecal P^H helps in the growth of lactose-fermenting organisms and suppresses the NH_3 forming bacterias (bacteroides).
5. It may detoxify short-chain fatty acids.

* Now-a-days, some clinicians use lactitol (30 g/day) instead of lactulose. Lactitol is available in powder form; it is less sweet, produces less flatulence and having quicker action.

Newer drugs in hepato-cellular failure :

- a) Acute—Flumazenil (benzodiazepine receptor antagonist), I.V branched-chain amino acid, levodopa, L-ornithine-L-aspartate (LOLA), sodium benzoate, probiotics.
- b) Chronic—Bromocriptine (dopamine agonist).

Drugs causing reduction of portal hypertension :

1. Vasopressin 2. Glypressin (terlipressin) 3. Somatostatin and octreotide 4. Propranolol (commonly used for prevention of recurrent bleeding) 5. Nitroglycerin 6. Nitrates, and 7. Verapamil.

Identification points for cirrhosis of liver :

1. Features of portal hypertension, and
2. Signs of hepato-cellular failure.

Always palpate the liver and spleen by 'dipping method' in the presence of ascites. *Splenomegaly is a very important bedside clinical finding to diagnose a case as cirrhosis of liver.* Never forget to examine for spider naevi, palmar erythema, gynaecomastia, scanty pubic hairs and testicular atrophy.

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Section 4

NERVOUS SYSTEM

The **cardinal symptoms of nervous system** comprise of :

(A) HIGHER FUNCTIONS :

1. Unconsciousness or alteration of consciousness, delirium, photophobia
2. Loss of memory; illusion, delusion or hallucination
3. Behavioural abnormality with speech defects

(B) CRANIAL NERVES :

1. Abnormal sensation of smell
2. Visual abnormality, diplopia (double vision), squint
3. Difficulty in mastication; loss of sensation over face
4. Deviation of angle of the mouth: facial asymmetry: epiphora
5. Loss of taste sensation
6. Vertigo, deafness, tinnitus
7. Nasal intonation or regurgitation
8. Hoarseness of voice
9. Dysphagia
10. Wasting of tongue, dysarthria

(C) MOTOR SYSTEM :

1. Loss of power or weakness, or clumsiness of movements*
2. Wasting / thinning, or swelling (pseudohypertrophy) of limbs
3. Fasciculation
4. Flexor spasms
5. Abnormal (involuntary) movements
6. Pain in the muscles
7. Unsteady gait

(D) SENSORY SYSTEM :

1. Anaesthesia
2. Paraesthesia
3. Tingling or numbness
4. Analgesia
5. Hyperaesthesia, hypoaesthesia
6. Root pain
7. Girdle-like sensation
8. Sensation of walking on cotton wool (e.g., tabes dorsalis)

(E) INCREASED INTRACRANIAL TENSION (IIT) :

1. Headache
2. Vomiting (projectile)
3. Convulsions
4. Blurred vision
5. Behavioural changes
6. Coma

(F) CEREBELLAR SYSTEM :

1. Swaying while walking, unsteadiness of gait or H/O falling (ataxia)
2. Weakness (as a result of hypotonia)
3. Difficulty in shaving, taking food to mouth and incoordination of speech
4. Tremor of hands while reaching for objects

(G) AUTONOMIC NERVOUS SYSTEM (INCLUDES BLADDER AND BOWEL FUNCTION) :

1. Retention of urine; feeling of sensation of bladder fullness
2. Incontinence (overflow or true)
3. Precipitancy
4. Hesitancy
5. Burning micturition [(signifies UTI); H/O catheterisation]
6. Impotence
7. Diarrhoea or constipation
8. Abnormal sweating (increased or diminished)

* For other features of autonomic nervous system, read the section on Trophic changes'

(H) MISCELLANEOUS :

1. Fever (meningitis, brain abscess)
2. Pain in the neck (meningitis, subarachnoid haemorrhage, cervical spondylosis)
3. H/O fall or seizure (epilepsy): H/O migraine; head injury; faintness (pre-syncope)
4. Psychological changes like depression or euphoria, agitation, sleep disorder
5. Discharge of pus from the ear (e.g., bacterial meningitis)
6. Bleeding tendency (e.g., thrombocytopenia)
7. H/O birth injury or anoxic injury

* Regarding **loss of power in the motor system**, enquire about involvement of :

- I. Upper limbs —
 - a) Proximal (lifting of arm above the head, combing, placing an object on a high shelf).
 - b) Distal (writing, sewing, typing or objects falling off the hand).
- II. Lower limbs —
 - a) Proximal (climbing upstairs and going downstairs, squatting and getting up from squatting position).
 - b) Distal (slipper falling from the feet).
- III. Trunk muscles—Ask the patient to sit up from supine position.
Moreover, ask about the ability of standing (with or without support), walking (with or without support), running etc. Predominant distal weakness is due to pyramidal tract involvement or peripheral neuropathy.

** Anaesthesia—loss of sensation, paraesthesia—tingling sensation (may be so intense as to be painful), analgesia—loss of pain sensation, hyperaesthesia—increased touch sensation, hyperalgesia—increased pain sensation, allodynia—perception of pain evoked by non-painful stimulus, hyperpathia—perception of pain that spreads out and outlasts the stimulus in time.

Scheme of Examination

- (A) *WHETHER THE PATIENT IS CONSCIOUS. ALERT AND CO-OPERATIVE :*
- (B) *HIGHER FUNCTIONS :*
1. Level of consciousness
 2. Appearance and behaviour
 3. Emotional state (euphoria, depression, hostile) i.e., the mood; sleep and dream.
 4. Orientation with time, place and person
 5. Illusion, delusion or hallucination
 6. Memory
 7. Intelligence
 8. **Speech** (with handedness)
- (C) *CRANIUM and SPINE :*
- (D) *NECK RIGIDITY. KERNIG'S SIGN. BRUDZINSKI'S SIGN. STRAIGHT LEG RAISING (SLR) TEST and EXAMINATION OF CAROTID ARTERIES :*
- (E) *CRANIAL NERVES :*
1. Olfactory—Test the smell sensation with common bedside objects like soap, toothpaste etc.
 2. Optic—
 - (i) Acuity of vision
 - (ii) Field of vision
 - (iii) Colour vision
 - (iv) Ophthalmoscopy or fundoscopy (not done)
 3. Oculomotor, trochlear and abducens—
 - (i) Ptosis
 - (ii) Squint
 - (iii) Enophthalmos or exophthalmos
 - (iv) Power of extraocular muscles—External ocular movements on follow and on command
 - (v) Nystagmus
 - (vi) Pupil— Size,
Shape,
Reaction (light reflex, consensual light reflex and accommodation reflex)
 4. Trigeminal—
 - (i) Motor function (masseter, pterygoids and temporalis)
 - (ii) Sensory function (sensation over the face)
 - (iii) Corneal reflex—Right and left side
 - (iv) Jaw jerk
 5. Facial—
 - (i) Palpebral fissure, frowning, symmetry of blinking, eye closure, nasolabial folds, angle of the mouth, blowing, whistling, showing the teeth, epiphora, dribbling of saliva
 - (ii) Power of individual facial muscle
 - (iii) Upper half of face escaped or not
 - (iv) Taste sensation of anterior 2/3rd of the tongue
 6. Vestibulocochlear—
 - (i) Hearing—Watch test, Rinne's test, Weber's test
 - (ii) Positional nystagmus (not done)
 7. Glossopharyngeal and vagus—
 - (i) Soft palate—Movements (on saying 'aah')
 - (ii) Pharyngeal or gag reflex
 - (iii) Taste sensation of posterior 1 /3rd of the tongue
 8. Spinal accessory—
 - (i) Power of sternomastoids
 - (ii) Power of trapezius
 9. Hypoglossal—
 - (i) Power of tongue muscles

- (li) Deviation
- (iii) Atrophy (size, shape, wasting)
- (iv) Fasciculation (or any abnormal movement)

(F) *MOTOR FUNCTIONS :*

a) Nutrition (inspection and palpation of muscles)—Attitude, atrophy or hypertrophy etc.

b) Tone— Right Left

(i) Upper limb : —

(ii) Lower limb : —

c) Power—

(i) Upper limb : —

(ii) Lower limb : —

d) Coordination—

(i) Upper limb : —

(ii) Lower limb : —

e) Involuntary movements —

(G) *SENSORY FUNCTIONS :*

a) Superficial or exteroceptive—

(i) Pain

(ii) Touch

(iii) Temperature (hot and cold)—not done; a rough assessment may be carried out at the bedside by using the metal of a tuning fork or metal part of stethoscope (cold perception) and by rubbing both palms (hot perception)

b) Deep or proprioceptive—

(i) Vibration sense

(ii) Muscle sense

(iii) Pressure sense

(iv) Joint sense

(v) Position sense

c) Cortical—

(i) One point localisation

(ii) Two point discrimination

(iii) Stereognosis

(iv) Graphaesthesia

(v) Sensory extinction (perceptual rivalry)

* Is there a definite line of demarcation of sensory loss in the trunk ?

** Both fine and crude touch sensations should be tested.

(H) *REFLEXES :*

a) Superficial— Right Left

1. Abdominal

(i) Upper —

(ii) Middle —

(iii) Lower —

2. Cremasteric —

3. PLANTAR RESPONSE —

b) Deep—

1. Biceps jerk —

2. Triceps jerk —

3. Supinator jerk —

4. Knee jerk —

5. Ankle jerk —

6. Clonus —

c) Visceral (sphincteric)—

1. Swallowing—The patient should be asked for dysphagia (with liquids, solids or both)

2. Bladder—The patient is asked regarding bladder and urethral sensation, retention, incontinence, urgency, or difficulty in controlling or initiating micturition

3. Bowel Whether facing any difficulty with defecation (constipation or incontinence). *Anal reflex* (gentle pricking of the skin on either side of the anus causes brisk contraction of the external anal sphincter which can be felt with a gloved finger at the anus; root value S S) may be tested^{3, 4}

- d) Other reflexes—
 1. Glabellar tap
 2. Grasp reflex
 3. Palmo-mental reflex

(I) *TROPHIC CHANGES* :

Bed sore, trophic ulcer, Charcot joint, changes in skin and hair

(J) *CEREBELLAR FUNCTIONS* :

Titubation, scanning speech, pendular knee jerk, intention tremor, finger-nose test, dysdiadochokinesia, nystagmus and reeling gait

(K) *AUTONOMIC FUNCTIONS* :

Temperature regulation, postural hypotension, impotence, abnormal sweating, nocturnal diarrhoea Horner's syndrome, trophic changes.

(L) *STANCE AND GAIT* (including Romberg's sign) :

* **Peripheral nerves** (this may be an added point in neurology) are examined for thickening and tenderness. Commonly examined peripheral nerves are :

- a) Great auricular nerve in neck
- b) Ulnar nerve at elbow
- c) Radial nerve at wrist
- d) Common peroneal nerve at the neck of fibula
- e) Sural nerve at the ankle

These nerves are commonly thickened in reference to leprosy. Read the causes of thickened peripheral nerves' from the section on 'Leprosy'.

Case 16

HEMIPLEGIA

What is your diagnosis ?

This is a case of left-sided complete hemiparesis (or hemiplegia) in the stage of recovery due to cerebral thrombosis probably involving the lenticulostriate branch of middle cerebral artery and the lesion is in the right internal capsule.

Why do you say so ?

This right-handed male patient aged 66 years was complaining of weakness of the left side of his body for last 15 days. At the onset when he was about to go to bathroom at about 5 a.m. he suddenly felt weak and could not move his left side. The episode was sudden in onset and the evolution of paralysis was completed within 6 hours. The patient complained of headache and vertigo at the beginning of the attack but neither he lost consciousness nor there was any difficulty in speech though there was some behavioural abnormalities present at that moment. He was immediately hospitalised. The patient also said that the weakness was marked in the left upper limb in comparison to left lower limb. There was no loss of bladder and bowel control. He had no H/O vomiting, convulsions, double vision, nasal regurgitation or nasal intonation; but he complained of deviation of face to the right side during eating, speaking or attempted smiling. Though the disease started acutely, he was gradually improving in the hospital.

There was no H/O fever, head injury, palpitation, chest pain, breathlessness, previous neurodeficit of this type which recovered completely (e.g., TIA); no family H/O diabetes mellitus, hypertension or tuberculosis was obtained. There was no H/O contact with tuberculosis, no H/O exposure to STD (sexually transmitted diseases). He did not give any H/O diabetes mellitus; a non-smoker by habit. There was no H/O waxing and waning. For the last few days he was on physiotherapy.

On examination, it shows that the patient is conscious and co-operative. He is hypertensive but neither there is any valvular heart disease present nor there is presence of any bruit over the carotids. He is suffering from UMN paralysis of left upper and lower limbs including the paralysis of lower half of left side of facial paralysis in the left) i.e., left-sided complete hemiparesis (or hemiplegia). The paralysis of left side of the body is suggested by the presence of clasp-knife spasticity, loss of superficial reflexes, brisk tendon reflexes and presence of extensor plantar response. The neurological examination of right side of the body was essentially normal.

What is your case ?

Say the summary as above.

Importance of 'past history' in hemiplegia :

1. Similar type of attacks or monoplegia in the past which recovered completely (indicates TLA).
2. Head injury (subdural haematoma).
3. H/O exposure to STD (neurosyphilis).
4. Hypertension (TLA, cerebral thrombosis or haemorrhage, lacunar infarction).
5. Diabetes mellitus (often associated with hypertension and accelerates the atherosclerosis).
6. Rheumatic fever (valvular heart disease and cerebral embolism).
7. Epilepsy (Todd's palsy) or migraine (neurologic migraine).
8. Tuberculosis (tuberculoma, tuberculous arteritis or tuberculous meningitis).
9. Intake of oral contraceptives with duration (may predispose to strokes in females, if used for more than 5 years), anticoagulants or aspirin; any bleeding tendency.
10. Fever (meningitis, cerebral abscess, encephalitis, leukaemia or lymphoma).
11. Dyspnoea or pain chest (mitral stenosis or recent acute myocardial infarction).
12. Intermittent claudication (atherosclerosis, arteritis).
13. Recent weight loss (tuberculosis).

* **Todd's palsy** — following epileptiform convulsions there may be development of paralysis (commonly monoplegia, rarely hemiplegia) due to exhaustion of cerebral neurones, and is known as Todd's palsy. Todd's palsy usually recovers within 24 hours.

Importance of 'family history' in hemiplegia :

1. Hypertension.
2. Diabetes mellitus.
3. Similar illness among other members of the family, specially in young patients (cerebral diplegia, spinocerebellar degeneration etc.)
4. Epilepsy or migraine.

Importance of 'personal history' in hemiplegia :

1. Obesity.
2. Sedentary habit.
3. Smoking.

Chief complaints (clinical presentations) of your patient :

1. Weakness of the left side of body for last 15 days,
2. Deviation of the face to right side while eating or speaking, for last 15 days, and
3. Difficulty in speech for last 15 days.

Enquiry regarding paralysis (in H/O present illness) :

(A) Date of onset : Note the specific date and time of onset of paralysis.

(B) Mode of onset : a) Sudden—Vascular, infective, traumatic and demyelinating
b) Gradual—Space occupying lesion (SOL).

N.B. ; (i) Cerebral embolism—develops in seconds (stormy).

- (ii) Cerebral thrombosis—develops in minutes and hours.
- (iii) Cerebral haemorrhage—very sudden in onset which relentlessly progress.
- (iv) Infective—develops in hours and days.
- (v) Demyelinating disease (multiple sclerosis)—develops in days and weeks.

(C) Precipitating factors :

- (i) During sleep or soon after rising from bed—Cerebral thrombosis.
- (ii) During exertion, activity—Cerebral embolism.
- (iii) At the height of emotion—Cerebral haemorrhage.
- (iv) During activity, exertion or height of emotion—Subarachnoid haemorrhage.
- (v) Following fever—Cerebral abscess, encephalitis, meningitis.
- (vi) Following convulsions—Todd's palsy.

(D) Evolution of paralysis :

- (i) Paralysis extends over hours to days ('stroke in evolution').
- (ii) Paralysis is 'total' or maximal from the beginning ('completed stroke')—Usually completed within 6 hours.

- (iii) Increase in a step-wise manner i.e., repeated episodes of TIA leading to fully evolved stroke ('stuttering stroke').

(E) Progress of paralysis :

- (i) Improving, stationary or deteriorating (a complete recovery usually indicates cerebral embolism or demyelinating disease).
 (ii) Waxing and waning (means exacerbations and remissions)—In demyelinating diseases.

(F) Degree and duration of paralysis : Weakness or total loss of power, with duration.

(G) Motor symptoms : See the symptomatology.

Meaning of few terminologies in clinical neurology :

'Plegia' means complete or near complete paralysis, and 'paresis' means weakness or partial paralysis.

1. Hemiplegia : Paralysis of one half of the body (specially of face, arm and leg) i.e., it is a facio-brachio-crural paralysis. The trunk is exempted due to bilateral innervation.
2. Hemiparesis : Weakness of one half of the body (specially of face, arm and leg).
3. Paraplegia : Paralysis of both the lower limbs (mostly due to spinal cord lesion).
4. Paraparesis : Weakness of both the lower limbs.
5. Diplegia : Bilateral hemiplegia of cortical origin where the lower limbs are more affected than the upper limbs.
6. Quadriplegia or tetraplegia : Paralysis of all the four limbs, and also the trunk (a feature of cervical cord disease).
7. Monoplegia : Paralysis of one limb—
 - (i) Brachial monoplegia—Paralysis of one upper extremity.
 - (ii) Crural monoplegia—Paralysis of one lower extremity.
8. Crossed hemiplegia : Paralysis of ipsilateral cranial nerves (LMN type) with contralateral hemiplegia (a feature of brainstem disease).
9. Cruciate hemiplegia : Brachial monoplegia of one side with contralateral crural monoplegia.
10. Bi-brachial paralysis : Paralysis of both upper extremities.
11. Bilateral hemiplegia : paralysis of all the four limbs, trunk, and total face.
12. Paraplegia with right brachial monoplegia : 3 limbs are involved (triplegia) i.e., paralysis of right upper limb and both the lower limbs.

Features of upper motor neurone (UMN) and lower motor neurone (LMN) lesion :

UMN Pyramidal cells (generally the Betz cells of area 4) and their axons upto the cranial nerve nuclei or anterior horn cells are called UMN (i.e., corticobulbar or corticonuclear, and corticospinal tracts).

LMN—The anterior horn cells and its dendrite upto the motor end plate, and cranial nerve nuclei with the cranial nerves constitute the LMN. LMN is the final common pathway for all motor impulses, both voluntary and automatic.

Table 8 : Features of UMN and LMN palsy

UMN palsy	LMN palsy
1. Muscle groups or limbs are affected	1. Paralysis of individual muscle supplied by that segment or nerve
2. Paralysis of voluntary movements*	2. Paralysis of muscles
3. Wasting is minimal and usually due to disuse atrophy	3. Atrophy and wasting are cardinal features
4. Clasp-knife spasticity (hypertonia)	4. Flaccidity (hypotonia)
5. Fasciculation is absent	5. Fasciculation may be present
6. Power is less affected than LMN type	6. Power is very much affected (may be Grade 0)
7. a) Superficial reflexes — Lost or diminished b) Deep reflexes or jerks — Brisk c) Clonus—May be present	7. a) Superficial reflexes—Lost b) Deep reflexes or jerks—Lost or diminished c) Clonus—Absent
8. Trophic changes in skin—rare	8. Trophic changes in skin—common
9. Plantar response—Extensor	9. Plantar response—Flexor or no response
10. Reaction of degeneration in the muscles—Absent	10. Reaction of degeneration—Present

* Actually, there is no paralysis of muscles in UMN palsy and it can be proved by the fact that the hemiplegic patient may run away from the room if the room suddenly catches fire.

What are 'strokes'?

The term 'stroke' is used to identify all forms of cerebrovascular accidents (CVA). A stroke or CVA is defined as 'focal neurological deficit of abrupt onset due to a vascular lesion, lasting longer than 24 hours, and is manifested, either as brain infarction or haemorrhage'.

Diseases under CVA i.e., types of CVA :

1. Cerebral thrombosis.
2. Cerebral embolism.
3. Cerebral haemorrhage.
4. Subarachnoid haemorrhage.
5. Hypertensive encephalopathy.
6. Others (inobvious stroke)—Multiple sclerosis, Todd's palsy, cerebellar haemorrhage, venous sinus thrombosis, subdural or epidural haematoma, cerebral metastasis etc.

N.B. : **Gradual onset hemiplegia** occurs due to :

1. Cerebral tumour.
2. Chronic subdural haematoma.
3. Cerebral abscess.
4. Meningitis and encephalitis.
5. AIDS (due to toxoplasmosis or primary CNS lymphoma).
6. General paralysis of insane (GPI).

* Stroke may be haemorrhagic or ischaemic. Cerebral thrombosis and cerebral embolism result in cerebral ischaemia, and ultimately lead to **cerebral infarction**. Cerebral vasculitis (e.g., SLE) may also give rise to cerebral infarction. Cerebral infarction is the commonest cause of hemiplegia in the elderly. Cerebral infarction literally means death of brain tissue.

** 'Stroke mimickers' are multiple sclerosis, hemiplegic migraine, cerebral tumour or abscess, and head injury.

*** Stroke is a better term than CVA.

What is lacunar infarction?

These are small, deep infarcts developed as a result of atherothrombotic or lipohyalinotic occlusion of perforating or penetrating arteries within the brain substance. The 'lacuna' (cavity after healing of infarction) measures 3 mm to 2 cm in diameter. The major risk factor remains hypertension. It may be present as 1) Pure motor hemiparesis, 2) Pure sensory stroke, 3) Ataxic hemiparesis, and 4) Clumsy hand, dysarthria syndrome.

Importance of examination of CVS in hemiplegia :

I. Method of examination :

1. Pulse—Rate, rhythm (regular or irregular), volume, condition of the arterial wall (thickened or not), comparison with opposite side, radio-femoral delay (whether there is any delay present or not, specially when the patient gives H/O intermittent claudication) and any special character.
2. Peripheral arterial pulses—
 - (i) **Carotids**—Kinking, diminished pulsation, or any bruit present over carotids or not.
 - (ii) Brachial—Locomotor brachialis (a feature of atherosclerosis).
 - (iii) Femoral,
 - (iv) Arteria dorsalis pedis, and
 - (v) Any bruit over the eyeballs (for arteriovenous malformation).
3. Blood pressure—High in cerebral haemorrhage and cerebral thrombosis (may be high in sub-arachnoid haemorrhage).
4. Jugular veins.
5. Apical impulse.
6. Heart sounds.
7. Murmur—Always search for a mitral diastolic murmur, specially in a patient with cerebral embolism.

II. Rheumatic heart disease—Cerebral embolism.

- III. Hypertensive heart disease—TLA, lacunar Infarction, cerebral thrombosis and haemorrhage.
- IV. Ischaemic heart disease—Mural thrombus may dislodge.
- V. Subclavian steal syndrome—The patient complains of dizziness, diplopia, ataxia and/or collapse after use of one arm (no neurodeficit is produced but one should search for difference between pulse and BP in two upper limbs, and a bruit over the suprascapular area), resulting from stenosis of subclavian artery proximal to origin of vertebral artery. The syndrome results from stolen blood from the brain via the vertebral artery when the said arm is in use.
- VI. Cyanotic congenital heart disease—May give rise to cerebral abscess, paradoxical embolism.
- VII. Syphilitic heart disease—Tabes dorsalis and GPI (not seen now-a-days).

Different sources in the heart for cerebral embolism :

- 1. Atrial fibrillation (from IHD, MS or MI)—Left atrial thrombus may dislodge.
- 2. SBE (always search for mild splenomegaly).
- 3. Left atrial myxoma.
- 4. Right-to-left shunt in the heart (paradoxical embolism in Fallot's tetralogy).
- 5. Left ventricular thrombus or mural thrombus in recent acute myocardial Infarction.
- 6. Left ventricular aneurysm.
- 7. Mitral valve prolapse.
- 8. Prosthetic heart valve.

Special points in the physical examination in hemiplegia :

(A) GENERAL SURVEY :

- a) The patient is conscious and co-operative.
- b) Facies—Angle of the mouth is deviated to the right side on attempted talking.
- c) Decubitus—Supine position in bed at present.
 - (i) Upper limb (left)—Flexed, adducted and semipronated; lying helplessly.
 - (ii) Lower limb (left)—Extended, adducted and plantiflexed; lying helplessly.
 - (iii) Right side (upper plus lower limbs)—Within normal limit.
- d) Neck veins—neither engorged nor pulsatile.
- e) Pulse—Mention all the points; condition of the arterial wall may be thickened.
- f) BP—It is compulsory to record the BP in all long cases; it is better to record BP on non-paralysed limb to avoid recording of low BP due to vasomotor paralysis on paralysed side; 185/ 100 mm of Hg In the present case.
- g) Respiration—Record rate, rhythm, type, depth and breathing pattern.
- h) Temperature—Record the axillary temperature (do not take oral temperature in facial palsy).
- i) Oedema—May present on the paralysed side due to prolonged immobility (1 venous + lymphatic flow) as well as affection of vasomotor nerves (reflex sympathetic dystrophy) .
- j) Any obvious deformity—Absent.

* Mention all the points in general survey.

(13) NERVOUS SYSTEM :

- a) The patient Is conscious, alert, oriented and co-operative.
- b) Higher functions including speech : Mention all the subheadings like level of consciousness, appearance and behaviour, memory etc.—NAD (no abnormality detected) or WNL (within normal limit); a bit dysarthric; right-handed person.
- c) Cranium and spine : Within normal limit.
- d) Neck rigidity, Kernig's sign : Absent; carotids—Within normal limit.
- e) Cranial nerves :
 - (i) VIIth cranial nerve—There is UMN type of paralysis on the left side of face as evidenced by,
 - Epihora (left).
 - Flattened nasolabial fold (left).
 - Deviated angle of the mouth (right) while showing teeth.
 - Weakness of buccinator, orbicularis oris, platysma (left).
 - Upper face on left side* (frontal belly of occipitofrontalis, corrugator superciliaris and upper part of orbicularis oculi) escaped paralysis.
 - (11) Other cranial nerves—Fundoscopy for papilloedema. Mention the subheadings under the different cranial nerves : NAD.



Football-like **distension of abdomen by ascites** in cirrhosis of liver. Venous prominence of portal hypertension is also evident; distance between xiphisternum to umbilicus is more than distance between umbilicus to symphysis pubis



Divarication of recti (bulging of linea alba in between two recti) and Maigne's bulgings (abdominal wall bulging above the iliac crests due to poor tone of external oblique muscles) seen on attempted 'rising test'



Bilateral pitting pedal **oedema** in congestive cardiac failure



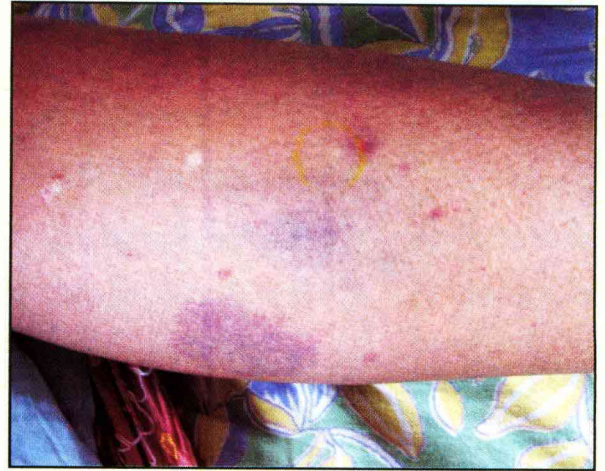
Parietal oedema demonstrated by pressing the tips of five fingers of right hand over the abdomen. Ascites and scar of past abdominal operation are also evident



Sacral oedema in a patient of congestive cardiac failure with prolonged recumbency. Here, the patient is sitting in the bed.



Haemorrhagic spots like **petechiae**, **purpura** and **ecchymoses** are visible over anterior chest and abdomen in septicemia



Shades of colours in **haemorrhagic spots** – bright red (fresh), blue (of some duration), and blackish-blue or black (old)



Herpes simplex virus infection (**herpes labialis**) leading to ulcerative stomatitis, in a patient recovering from meningitis



Purple striae developed after long-continued corticosteroid therapy



White striae noted after repeated paracentesis abdominis in ascites

f) Motor functions :

	Right	Left
(i) Nutrition	Normal. No wasting seen	Normal plus write down the attitude of left side from the 'Decubitus'. No wasting seen
(ii) Tone	a) Upper limb—Normal b) Lower limb—Normal	Clasp-knife spasticity in both the limbs. Flexor tone is increased more in upper limb and extensor tone is increased more in lower limb
(iii) Power	a) Upper limb—Grade V b) Lower limb—Grade V	Grade III in both the limbs
(iv) Coordination	a) Upper limb—Normal b) Lower limb—Normal	Could not be tested properly
(v) Involuntary movements	None	None

g) Sensory functions :

(i) Superficial	N	
(ii) Deep	I	NAD
(iii) Cortical	J	

* Mention the subheadings.

h) Reflexes :

(i) Superficial—	
a) Abdominal—	
x) Upper ■	
y) Middle	► Normal Lost
z) Lower ,	
b) Cremasteric	Normal Lost
c) Plantar response	Flexor Extensor
(ii) Deep—	
a) Biceps jerk	
b) Triceps jerk	
c) Supinator jerk	Normal Brisk
d) Knee jerk	►
e) Ankle jerk	
f) Clonus	Absent Absent

(say, if present)

(iii) Visceral—

a) Bladder I	
b) Bowel	Within normal limit
c) Swallowing J	

(iv) Other reflexes—

a) Glabellar tap	Normal
b) Grasp reflex	Absent
c) Palmo-mental reflex	Absent

i) Trophic changes : Absent.

j) Cerebellar functions : Within normal limit,

k) Autonomic functions : Normal.

l) Gait : Classical hemiplegic gait. The patient walks by tilting the pelvis while the left leg is making 'circumduction'. Read the section on 'Abnormal gait'.

(C) CVS : See the answer of the question 'Importance of examination of CVS in hemiplegia' mentioned earlier (write the points from 1 to 7 under I).

(D) RESPIRATORY SYSTEM :

- (i) Shape of the chest
- (ii) Movement of the chest
- (iii) Trachea and apex beat
- (iv) Percussion—normal resonant note on both sides.
- (v) Breath sound—vesicular.
- (vi) Vocal resonance—normal on both sides.
- (vii) Adventitious sound—none.

} within normal limit

(E) G.I TRACT AND GENITOURINARY SYSTEM :

- (i) Mouth, tongue, teeth, gum — NAD.
- (ii) Shape of the abdomen — NAD.
- (iii) Umbilicus — Inverted.
- (iv) Rigidity — Absent.
- (v) Hepatosplenomegaly — Not present.
- (vi) Any lump felt — None.
- (vii) Fluid thrill and shifting dullness—Absent. Percussion for bladder fullness done.
- (viii) Auscultation — Normal peristaltic sound.
- (ix) Genitalia — NAD
- (x) P/R examination—not done.

(F) LYMPHORETICULAR SYSTEM :

- (i) Lymphadenopathy — Absent.
- (ii) Sternal tenderness — Absent.
- (iii) Haemorrhagic spots — Absent.

Clinical types of hemiplegia :

1. Complete hemiplegia (with UMN type Vllth nerve palsy in the side of hemiplegia).
2. Incomplete hemiplegia (hemiplegia without affection of Vllth cranial nerve).
3. Crossed hemiplegia (when the site of lesion is in the brainstem) Ipsilateral LMN type cranial nerve paralysis (i.e., at the side of brainstem lesion) with contralateral hemiplegia.

Importance of headache, convulsions and coma (features of IIT) in strokes :**(A) HEADACHE—**

- (i) Severe headache accompanying the paralysis—Subarachnoid haemorrhage (occipital headache), cerebral haemorrhage, hypertensive encephalopathy.
- (ii) Headache preceding the paralysis for months—SOL (space occupying lesion), subdural haematoma.
- (iii) Mild headache—cerebral thrombosis or embolism.

(B) CONVULSIONS—

- (i) Generalised convulsions preceding the paralysis—Todd's palsy (post-epileptic), hypertensive encephalopathy or SOL.
- (ii) Focal or generalised convulsions occurring few months prior to paralysis — SOL.
- (iii) Convulsions just prior to the paralysis may be seen in cerebral haemorrhage, thrombosis or embolism. Convulsions do not occur in subarachnoid haemorrhage.

(C) UNCONSCIOUSNESS—

1. From the beginning—Commonly cerebral haemorrhage.
2. In cerebral thrombosis, cerebral embolism, subarachnoid haemorrhage or hypertensive encephalopathy—Usually the patients remain conscious but may be drowsy.

What is projectile vomiting of increased intracranial tension (IIT) ?

It is the sudden vomiting which is not preceded by nausea (common in children).

Enquiry regarding the bladder and bowel symptoms in neurology :

Following questions are put to a hemiplegic or paraplegic patient :

1. Is there any increased frequency or urgency ?
2. Do you feel burning sensation during micturition ?
3. Is there any problem in initiation of micturition (hesitancy or precipitancy) ?

4. Can you feel the urine passing ?
5. Can you stop the flow of urine in the midstream at will ?
6. Is there any dribbling of urine ?
7. Is there any sense of bladder fullness ?
8. Is there any associated rectal disorder (incontinence of stool, diarrhoea or constipation etc) ? or any difficulty in anal sensation ?
9. Is there any disorder of potency (in males) ?
10. Have you ever catheterised in recent past ?

N.B. 1, 2, 3 and 10 are important in hemiplegia. In paraplegia, all the points are important.

Type of bladder involvement in your patient :

The chief complaint of the patient is burning micturition at present (seen in many cases) as he was catheterised on admission (and now suffering from UTI). There is no loss of bladder control.

Point out the signs of UMN lesion in your patient :

Demonstrate the points mentioned below :

1. Vllth cranial nerve—UMN palsy (as upper face on the left side escaped paralysis).
 2. Power—Loss of power.
 3. Tone—Clasp-knife spasticity (hypertonia).
 4. Reflexes—
 - a) Abdominal—Lost.
 - b) Jerks—Brisk.
 5. **Plantar response—Extensor.**
- All the features are left-sided in this patient. Add clonus, if present.

Why there is Vllth nerve palsy (UMN type) in hemiplegia ?

Remember the principle : The Vllth cranial nerve nuclei are divided into upper and lower part by an arbitrary horizontal line, and the upper part supplies muscles of upper face while the lower part supplies the lower face. All cranial nerve nuclei are bilaterally supplied by pyramidal fibres (corticonuclear fibres) except the lower part of the Vllth cranial nerve nuclei which receive pyramidal fibres only from the opposite side. So the lower face is easily affected (i.e.. UMN type of Vllth nerve palsy) in a patient of hemiplegia.

Trunk and abdominal muscles in hemiplegia :

These are not involved in classical hemiplegia (lesion at internal capsule) due to their bilateral representation at the cortex.

- Abdominal muscles ($T_5 - L_1$) : Do the 'rising test' and analyse "Beevor's sign".
- Intercostal muscles ($T_1 - T_{12}$) : During respiration, observe the movements of intercostals as well as the ribs.
- Extensors of spine (all segments of spinal nerve) : Patient lies prone and tries to raise his shoulder from the bed against examiner's resistance. Back muscles (e.g., erector spinae) are seen to contract.

Stages of hemiplegia and their clinical diagnosis :

1. Stage of neural shock (for few hours to days, and may extend upto 2-3 weeks).
2. Stage of recovery.
3. Stage of residual paralysis.

Diagnosis :

(A) STAGE OF NEURAL SHOCK (a stage when all reflex activity is suppressed) :

- (i) Patient may be drowsy or in coma.
- (ii) Aphasia.
- (iii) Hypotonia of muscles or flaccidity with gross diminution of power.
- (iv) Jerks are usually absent.
- (v) Retention of urine and there may be spontaneous evacuation of faeces.
- (vi) Plantar response—No response or equivocal.

(B) STAGE OF RECOVERY :

Recovery usually occurs in the following order :

- (i) Face earliest.

- (ii) Lower limb (first the extensors recover).
- (iii) Upper limb (first the flexors recover).
- (iv) Dorsiflexion of foot (minimally recovered).
- (v) Finer movements of fingers (usually do not recover).

(C) STAGE OF RESIDUAL PARALYSIS :

Patient lives with some residual paralysis and walks in classical hemiplegic gait. During recovery of UMN lesion reflex recovers first, then comes tone and lastly power (proximal earlier than distal power). Plantar response may remain extensor on the affected side throughout life.

Importance of examination of cranial nerves in hemiplegia :

It helps in localisation of the site of lesion. The olfactory and optic nerves are actually built up of central nervous tissue rather than peripheral nerves. The disposition of other cranial nerve nuclei in the brainstem are as follows :

- a) Midbrain — IIIrd and IVth.
- b) Pons — Vth, VIth, VIIth and VIIIth.
- c) Medulla — IXth, Xth, XIth and XIIth.

Different 'brainstem syndromes' are :

1. Weber's syndrome—Paralysis of IIIrd cranial nerve of one side (LMN type) with contralateral hemiplegia (crossed hemiplegia)—lesion is at midbrain.
2. Millard-Gubler syndrome—Paralysis of VIth cranial nerve (LMN type) ± VIIth cranial nerve palsy (LMN type) with contralateral hemiplegia (crossed hemiplegia)—lesion is at pons.
3. Avellis syndrome—Paralysis of Xth cranial nerve of one side (LMN type) with contralateral hemiplegia (crossed hemiplegia)—lesion is at medulla.

* *Crossed hemiplegia always indicates the lesion in the brainstem.*

Importance of VIIth cranial nerve examination in hemiplegia :

It helps in localisation of the site of lesion in hemiplegia. For example :

- a) UMN type of VIIth cranial nerve palsy in the side of hemiplegia (as this case is)—Lesion is 'above' the pons (opposite side).
- b) LMN type of VIIth cranial nerve palsy with contralateral hemiplegia i.e., crossed hemiplegia (lesion is at VIIth cranial nerve nucleus)—Lesion is 'at' pons on the side of VIIth nerve palsy.
- c) No facial nerve involvement (incomplete hemiplegia)—Lesion is below the level of pons.

* ***UMN type of VIIth cranial nerve palsy is always associated with same-sided hemiplegia while hemiplegia, if at all present with LMN type of VIIth cranial nerve palsy, is always crossed.***

Eye position in hemiplegia :

- (A) Hemisphere lesion : eyes deviated towards the side of lesion in cerebral hemisphere.
- (B) Pontine lesion : eyes deviated to the paralysed side.
- (C) Putamen lesion : loss of conjugate lateral gaze.
- (D) Thalamic lesion : loss of upward gaze, down or skew deviation, lateral gaze palsies, unequal pupil.

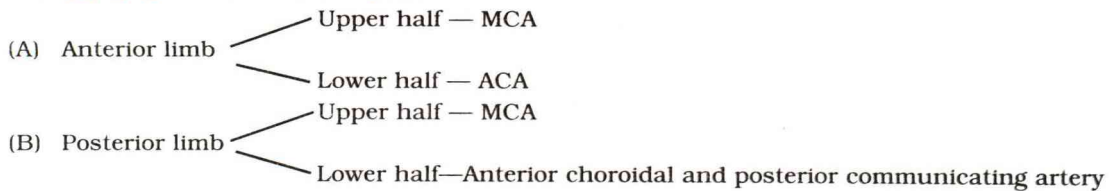
Functions of pyramidal and extrapyramidal system :

- (A) Pyramidal tract—Helps in volitional movements, specially the skillful movements (movement of fingers and hands e.g., writing, buttoning, playing piano etc).
- (B) Extrapyramidal tract—
 - (i) Maintains muscle tone and posture.
 - (ii) Maintains emotional, automatic and associated movements (i.e., swinging of the hands during walking).
 - (iii) Checks abnormal involuntary movements.

Blood supply of the brain in a nutshell :

- (A) Anterior cerebral artery (ACA)—Supplies branches to frontal and parietal lobes, to part of internal capsule; it also supplies the paracentral lobule i.e., the centre for movements of the lower limbs plus the centre for micturition and defecation.
- (B) Middle cerebral artery (MCA)—Supplies the motor, sensory and speech areas.
- (C) Posterior cerebral artery (PCA)—Supplies the visual cortex, occipital lobe and part of the temporal lobe; part of the posterior limb of the internal capsule.

Blood supply of the internal capsule :



N.B. : Pyramidal fibres take origin from Betz cells of cerebral cortex and descend towards corona radiata. While descending from corona radiata, pyramidal tract occupies the anterior $\frac{2}{3}$ rd of the posterior limb of the internal capsule and genu. Anterior limb contains the fibres for the face while fibres for the tongue and mouth lies at the genu. Pyramidal fibres occupying the anterior $\frac{2}{3}$ rd of the posterior limb continue downwards and occupy the middle $\frac{3}{8}$ th of the peduncles of midbrain. Then the fibres descend through pons and medulla, and majority of the fibres cross to the opposite side to occupy the lateral column in the spinal cord.

* As pyramidal tract has contralateral supply, hemiplegia occurs contralateral to the side of lesion in brain.

Features of anterior cerebral artery involvement :

If it is blocked 'distal' to the anterior communicating artery, there is :

1. Hemiplegia and hemisensory loss of the opposite side of body where lower limbs are much more involved (crural dominance) than upper limbs (in hemiplegia due to blocked middle cerebral artery, the weakness is more marked in upper limbs i.e., there is brachial dominance).
2. Face is not involved.
3. Urinary incontinence is very common.
4. Presence of mental symptoms (emotional disturbance).
5. Presence of 'released reflexes'.

* If anterior cerebral artery is blocked 'proximal' to anterior communicating artery, there is development of complete hemiplegia (hemiplegia + face involvement) on the opposite side with or without aphasia.

** In Heubner's artery involvement (a branch of anterior cerebral artery), opposite sided facio-brachial (face + hand) paralysis occurs.

Possible sites of lesion in hemiplegia :

1. Cortical and subcortical.
2. Internal capsule.
3. Brainstem.
4. Spinal.

Characteristics of each site of lesion :

(A) CORTICAL :

1. As the fibres are widely distributed, an extensive lesion (outcome is obviously grave) is necessary to produce contralateral hemiplegia. Usually cortical lesion produces **monoplegia**.
2. Jacksonian convulsions and headache are common at the onset. Speech abnormality is present.
3. Flexors and extensors of both the upper and the lower limbs are equally involved, and so the involvement of tone of muscles.
4. There may be cortical type of sensory loss (e.g., astereognosis).

(B) SUBCORTICAL (OR AT THE LEVEL OF CORONA RADIATA) :

1. Subcortical lesion involves more fibres than the cortical lesion of equal size. Usually it produces **monoplegia** but hemiplegia on the opposite side may occur. There is absence of cortical signs like convulsions or dysphasia.
2. Involvement of adjacent thalamocortical fibres result in impairment of postural sensibility, tactile localisation and discrimination in the affected limbs.

(C) INTERNAL CAPSULE :

1. It is the most common site of lesion in a CVA.
2. As the fibres are densely packed, a small lesion may produce **complete hemiplegia on the opposite side**.

3. Hemianaesthesia and homonymous hemianopia occur as a result of damage to the sensory and the visual fibres respectively which lie posterior to the pyramidal tract at internal capsule.
4. Global aphasia (in left-sided lesion) or * apractagnosia (in right-sided lesion).
5. Flexor tone is more in upper limb and the extensor tone is more in lower limb (in hemiplegic side). Classical position of the hemiplegic limb :
 - a) Upper limb—Flexed (elbow, wrist and fingers), adducted and semipronated.
 - b) Lower limb—Extended, adducted and plantiflexed.
6. Cranial nerve involvement—Only UMN type of the VIIth cranial nerve involvement on the side of paralysis, as discussed earlier.

* A type of agnosia marked by inability to use objects or perform skilled motor activities.

(D) BRAINSTEM :

1. Dizziness, vertigo and vomiting may be present.
2. Always **crossed hemiplegia**.
3. See the different brainstem syndromes discussed earlier and localise the site of lesion.
4. Gaze paralysis.
5. Change in pupil is common, even Horner's syndrome may be seen.
6. Involvement of the cerebellum may be there.

(E) SPINAL HEMIPLEGIA :

Though rare, lesion lying below the medulla but above the C₅ spinal segment may produce '**ipsilateral**' hemiplegia (as the pyramidal fibres have already crossed in medulla) without any cranial nerve involvement (i.e., incomplete hemiplegia).

Classical features of cerebral thrombosis, embolism, haemorrhage etc :

(A) CEREBRAL THROMBOSIS :

1. Middle aged or elderly; commonest cause of CVA.
2. Very often hypertensive and atherosclerotic.
3. Onset is less rapid than cerebral embolism or haemorrhage; usually with step-wise progression.
4. Mild headache and convulsions may occur. There may be transient loss of consciousness.
5. Often there is past history of TLA (single or multiple).
6. Occurs during sleep or soon after rising from bed.
7. Carotid bruit with diminished pulsation highly supports cerebral thrombosis.

(B) CEREBRAL EMBOLISM :

1. Commonly in young females.
2. Valvular heart disease or sources of embolism (discussed earlier) may be evident.
3. Pulse may be irregularly irregular (atrial fibrillation).
4. Develops in seconds (dramatic, stromy or 'bolt from the blue') during exertion, activity or any time; warning sign of TIA may be present.
5. Headache and convulsions are usually absent; there may be transient loss of consciousness.
6. Shifting hemiplegia may occur. Recovery may be very rapid.
7. Evidence of systemic embolisation may be present.

(C) CEREBRAL HAEMORRHAGE :

1. Usually above the age of 40 years.
2. Develops acutely at the height of emotion or excitement. Develops over minutes and hours which progresses relentlessly.
3. Almost all are hypertensive.
4. Headache, recurrent vomiting or convulsions are present in most of the patients. Unconsciousness gradually progresses to deep coma. Early recovery is unusual.
5. Heart may show features of LVH.
6. Features of atherosclerosis may be present (as in thrombosis).
7. Conjugate deviation of eyes and Cheyne-Stokes breathing are common.
8. Very rarely, neck rigidity may be elicited.

* Lenticulostriate branch of MCA is commonly involved in hypertensive haemorrhage.

** Features of pontine haemorrhage are pin-point pupil, pyrexia and paralysis (3P).

*** Common sites of intracerebral haemorrhage in order of frequency are : Putamen -> thalamus -> frontal or parietal lobes -> pons -> cerebellum.

(D) SUBARACHNOID HAEMORRHAGE :

1. Usually in young adults (also known as 'apoplexy of the young'). It may occur in older age group. Subarachnoid haemorrhage occurs as a result of rupture of berry aneurysm or arterio-venous malformations.
2. Bursting occipital headache commonly precedes (often called 'as the worst headache of my life'); usually does not give rise to convulsions but loss of consciousness (50%) may be present.
3. Occurs during activity or at the height of emotion.
4. Neck rigidity and Kernig's sign are present (cardinal signs at bedside).
5. Hemiplegia or aphasia is absent in most of the patients.
6. Sometimes, there is paralysis of IIIrd, IVth, VIth cranial nerves, or monoplegia or hemiplegia (due to local pressure effect of berry aneurysm or vascular spasm).

(E) CAROTID HEMIPLEGIA :

1. Middle aged or elderly.
2. Very often hypertensive and atherosclerotic.
3. History of TIA is commonly found.
4. There may be H/O claudication of the jaw.
5. Kinking of the carotid artery, inequality of the pulse volume in two carotids or a bruit may be heard at the site of the block.
6. Ipsilateral visual disturbances (amaurosis fugax—transient mono-ocular blindness) and contralateral sensory-motor deficit.

* HYPERTENSIVE ENCEPHALOPATHY is an acute syndrome where severe hypertension is associated with nausea, vomiting, excruciating headache, transient abnormalities in speech or vision, convulsions, mental confusion and coma. Focal neurological signs are rarely seen but papilloedema is usually present. The symptom complex results from constriction of cerebral arterioles and cerebral oedema. Neurodeficit is fully correctable if acute hypertension is properly controlled.

** Following events are **common in hypertensive patients** like cerebral haemorrhage or thrombosis, TIA, carotid atheroma, subarachnoid haemorrhage and hypertensive encephalopathy.

Features of 'atherosclerosis' :

1. Age of the patient is usually above 50 years.
2. There may be H/O IHD, TIA, intermittent claudication or hypertension.
3. Risk factors like diabetes mellitus, smoking, hyperlipidaemia, central obesity may be present.
4. Condition of the arterial wall—Thickened (arteriosclerosis), feels like a cord and often tortuous (Monckeberg's medial sclerosis), and may be associated with :
 - (i) Locomotor brachialis.
 - (ii) Tortuous temporal arteries, and
 - (iii) Kinked carotids.
6. There may be presence of suprasternal pulsation.
7. Features of hyperlipidaemia like xanthelasma is seen around the eyes.
8. Often associated with corneal 'arcus' and diagonal ear lobe crease.
9. Change of arterial and venous ratio, and arteriovenous nipping in ophthalmoscopy.
10. Chest X-ray may reveal unfolding of aorta or aortic calcification.

Risk factors in CVA :

- | | |
|---|---|
| 1. Systemic hypertension. | 8. Oral contraceptive pills. |
| 2. Diabetes mellitus. | 9- High alcohol consumption. |
| 3. Hyperlipidaemia. | 10. Hyperviscosity syndrome (polycythemia, thrombocythemia etc.), bleeding diathesis. |
| 4. Smoking. | H- Trauma. |
| 5. Obesity. | 12. Strong family history (heredity). |
| 6. Cardiac diseases (valvular and ischaemic). | 13. Sleep apnoea. |
| 7. Severe carotid stenosis. | |

* Increased age, male sex (except in extremes of ages) and hyperhomocysteinaemia are other risk factors.

** Major risk factors : 1- 7, and 10; minor risk factors : 8, 9 and 11-13.

Common causes of 'strokes in young' (<40 years of age) :*(A) CAUSES :*

1. Cerebral embolism (from cardiac sources like rheumatic heart diseases; vide page 142).

2. Subarachnoid haemorrhage (rupture of berry aneurysm, arteriovenous malformations).
3. Arteritis or vasculitis (tuberculosis, syphilis, Takayasu's disease, collagen vascular diseases).
4. Accelerated atherosclerosis (secondary hypertension, familial hypercholesterolaemia, hypertriglyceridaemia, diabetes mellitus, nephrotic syndrome, SLE).
5. Hyperviscosity syndrome (polycythemia, thrombocytosis).
6. Demyelinating disease (multiple sclerosis).
7. Intracranial neoplasm (primary or secondary).
8. Encephalitis.
9. Head injury (trauma).
10. Anticoagulant therapy, ITP. or treatment with oral contraceptive pills.
11. Cerebral abscess, tuberculoma.
12. Hemiplegic migraine.
13. 'Procoagulant states' e.g., Protein C and S deficiency, antithrombin III deficiency, hyperhomocystinaemia, activated protein C resistance, factor V Leiden, prothrombin mutations, antiphospholipid syndrome (Hughes syndrome).
14. Venous sinus thrombosis—Specially in the setting of puerperium.
15. Cerebral malaria.
16. Drug abuse (e.g., cocaine).

(B) SPECIAL INVESTIGATIONS IN YOUNG PATIENTS :

- a) Full blood count will platelet. ESR, VDRL test, sugar, urea; X-ray, ECG and echocardiography.
- b) Antinuclear factor (ANF); antibodies to double-stranded DNA.
- c) Anti-cardiolipin antibodies (antiphospholipid syndrome may be a cause of stroke in young patients), protein C and S, antithrombin III.
- d) Prothrombin time (PT), activated partial thromboplastin time (aPTT).
- e) Lupus anticoagulant.
- f) Lipid profile.

* Atherosclerosis, the chief cause of cerebral ischaemia in the aged is seen less commonly in young patients with stroke. The causes of 'strokes in young' are a bit different from the aged population.

Possible causes of thrombo-embolism :

- (A) Arterial : systemic hypertension, infected or sterile emboli of cardiac or vascular origin, septicaemia, hyperhomocystinaemia, thrombotic thrombocytopenic purpura, myxoma, Takayasu's arteritis, polyarteritis nodosa.
- (B) Venous : venous insufficiency/obstruction, prolonged immobility, nephrotic syndrome, antiphospholipid syndrome, oral contraceptive pills, plus the 'procoagulant states' for both the categories (described above).

Causes of recurrent / transient hemiplegia :

1. Transient ischaemic attack (TIA).
2. Post-epileptic or Todd's palsy.
3. Hypertensive encephalopathy.
4. Hemiplegic migraine.
5. Multiple sclerosis.
6. Hysterical hemiplegia.

* 'Reversible' neurodeficit is common in 1, 2, 4 and 6.

Major side effects of prolonged use of oral contraceptive pills :

1. Obesity or weight gain.
2. CVA (thrombo-embolic strokes commonly).
3. Deep vein thrombosis and pulmonary embolism.
4. Hypertension.
5. Increased Incidence of acute myocardial infarction.
6. Cholestatic jaundice.
7. Increased incidence of cholelithiasis and cholecystitis.
8. Impairment of glucose tolerance and worsened lipid profile.
9. Depression.
10. Breast discomfort, chloasma, increased clotting tendency, migraine, loss of libido, drug interactions (e.g., rifampicin) and abnormal liver biochemistry.

* Recently, in OC pills, low dose oestrogens have been combined with newer progestational agents, which cause less deleterious effects on carbohydrate and lipid metabolism, coagulation profile, vascular apparatus and liver function.

Mention few causes of monoplegia :

(A) BRACHIAL MONOPLÉGIA (UPPER LIMB) :

1. CVA (cortical or subcortical lesion).
2. Multiple sclerosis.
3. Encephalitis.
4. Poliomyelitis.
5. Mononeuropathy.
6. Brachial plexus lesion.
7. Motor neurone disease (MND)
8. Hysterical.

(B) CRURAL MONOPLÉGIA (LOWER LIMB)

1. Thrombosis of paracentral artery.
2. Multiple sclerosis.
3. Cauda equina lesion.
4. Sciatic nerve palsy.
5. Prolapsed intervertebral disc (PID).
6. Mononeuropathy.
7. Hysterical.

Causes of hemiplegia :

1. Cerebrovascular accidents (page 141).
2. Strokes in young (page 149).
3. Gradual onset hemiplegia (page 141).
4. Recurrent and reversible hemiplegia (page 150).

Actually, these are sumtotal causes of hemiplegia.

Why do you consider cerebral thrombosis in this case ?

1. As the onset was dramatic, I think the pathology is a 'vascular catastrophe'.
2. Though there is presence of hypertension, neither the patient gave any H/O vomiting and convulsions at onset nor he was unconscious. So I am not thinking of cerebral haemorrhage in this case. As there is no neck rigidity, no H/O bursting occipital headache but there is presence of hemiplegia, I am not thinking of subarachnoid haemorrhage in this patient.
3. The CVS is within normal limit. The patient is not suffering from any valvular heart disease or atrial fibrillation. So chance of cerebral embolism is less.
4. So, I say it is a case of CVA due to thrombosis which is the 'most common' cause of stroke.

Why do you say the site of lesion is in the internal capsule ?

It is known that in a patient of hemiplegia if there is,

1. UMN type of VIIth nerve palsy to the side of hemiplegia—the lesion lies in the opposite internal capsule.
2. LMN type of VIIth nerve palsy with contralateral hemiplegia—the lesion lies in the brain stem (pons) opposite to hemiplegic site.
3. Hemiplegia without any cranial nerve involvement (known as spinal hemiplegia) lesion lies in ipsilateral upper spinal cord (in between medulla to C₅ spinal segment).

As it is a case of left-sided complete hemiparesis, most likely the site of lesion is in the right internal capsule. It is the 'commonest site' of involvement in a patient with CVA.

* Cortical and subcortical lesions usually produce monoplegia.

'Clinical' classification of stroke :

1. TIA (transient-ischaemic attack)—Abrupt neurological dysfunction (neurodeficit) due to cerebral ischaemia where symptoms last for < 24 hours and the patient recovers completely.
2. Stroke in evolution (progressive stroke)—The symptoms worsen gradually or in a step-wise pattern over hours or days. This is due to increasing size of infarction, haemorrhage or cerebral oedema.
3. Completed stroke—The neurodeficit is complete from the beginning (usually within 6 hours) and persists for weeks and months, and often permanently.
4. Minor stroke—Usually the patient recovers within a week, without significant deficit.

* Most neurologists still believe that TIA is a stroke; the definition of TIA is unsatisfactory as clinical studies reveal that after J hour, ischaemic damage occurs. TIA of (a) *carotid territory* manifests as transient weakness (monoparesis), hemisensory loss, aphasia and transient mono-ocular blindness (amaurosis fugax) whereas, (b) *vertebro-basilar TIA* is manifested by vertigo, dysarthria, diplopia, vomiting,

ataxia, dysphagia or sudden fall (drop attack). In TIA, the source of embolus may be clinically evidenced by carotid arterial bruit (i.e., stenosis), valvular heart disease/SBE, atrial fibrillation, recent acute myocardial infarction or difference between BP of left and right arm.

The older term RIND (reversible ischaemic neurological deficit) where the neurodeficit persists for > 24 hours but recovers usually within 24-96 hours, is not preferred by neurologists now-a-days.

Classical 'signs of meningeal irritation' :

1. Neck rigidity (stiffness)—

- a) In *supine position* of the patient, stand on the right side of the bed and place your left hand behind and below the patient's head; try to lift the head from the bed until the chin touches the chest and feel the resistance while flexing the head. Normally, the neck is supple and can be easily flexed passively. In the presence of neck rigidity, it is not possible to passively flex the neck fully. Neck rigidity occurs due to spasm of extensor neck muscles and in an attempt to do the test, the whole body may be raised up; head retraction represents an extreme form of neck rigidity (alternative method : support the back of the head with both hands and gently try to flex the neck so that the chin touches the chest).
- b) In *sitting position* of the patient, ask him to touch the chin to the sternum (always with closed mouth). Neck rigidity and Kernig's sign are present in,

(i) Meningitis.	(vii) Rarely in intracerebral haemorrhage.
(ii) Meningism.	(viii) Spinal epidural abscess.
(iii) Subarachnoid haemorrhage.	(ix) Sometimes in posterior fossa tumour.
(iv) Meningoencephalitis.	(x) Cerebellar tonsillar herniation at foramen magnum.
(v) Meningomyelitis.	
(vi) Tetanus, rabies, hysteria.	

* In clinical practice, No. 1, 2, and 3 are most important.

N.B. : Sometimes, the *neck rigidity is absent* in deep coma or in infants. Passive resistance to head flexion may be present in cervical spondylosis (side to side movement is more restricted than antero-posterior movement), traumatic injury of cervical spine, Klippel-Feil anomaly or cerebral malaria.

2. **Kernig's sign**—In supine position of the patient, flex his hip to 90° and then extend the knee to 90°. Now try to straighten the leg maintaining the hip in flexion. In the presence of meningeal irritation, it is impossible to straighten the leg because of the painful spasm of hamstring muscles (normally the leg can be easily straightened). This sign is positive in the conditions mentioned above. Testing of neck rigidity is a more sensitive test of meningeal irritation than Kernig's sign.

3. Brudzinski's sign—

It is a very *helpful sign of meningeal irritation in children*. It has two components :

- a) Neck sign—Patient will lie supine with extended legs. Try to lift the head from the bed. There will be reflex flexion of the hip, or knee of one or both the limbs.
- b) Leg sign— While passively flexing the thigh of any one limb, the other thigh or knee automatically becomes flexed.

* Kernig's and Brudzinski's sign are due to the presence of inflammatory exudate around roots present in the lumbar theca, which results in muscle spasm.

How to differentiate meningism from meningitis ?

Presence of neck rigidity and rise in CSF pressure are found in both the conditions but,

1. If along with neck rigidity, there is presence of pain in the neck — Probably it is meningitis (sign of inflammation). Meningism usually does not produce pain.
2. Kernig's sign is less pronounced in meningism.
3. Convulsions and coma are rare in meningism, and cranial nerve paralysis never occurs.
4. If after lumbar puncture, CSF is seen to be turbid — It is meningitis. Clear CSF may be seen both in meningism and meningitis (viral or tuberculous).
5. CSF shows absence of cellular changes in meningism. and the fluid is always sterile on culture. Meningitis will always have changes in cell count and cell type.

Diseases producing meningism in clinical practice :

- | | |
|---|---|
| 1. Enteric fever. | 5 Acute pyogenic tonsillitis. |
| 2. Pneumonia (commonly atypical variety). | 6. Viral encephalitis; any viral infection. |
| 3. Empyema thoracis. | 7 Leukaemia, lymphoma, malignancy. |
| 4. Diphtheria. | g Weil's disease. |

* *Meningism (meningeal irritation)* is usually due to some local or systemic infection but there is absence of any direct infection or inflammation of CNS.

What is straight leg raising (SLR) test ?

While the patient lies supine, try to elevate patient's 'extended' leg, keeping the hand behind the patient's heel. This movement is possible upto 90° in a normal person. The test is positive i.e., restricted movement with pain is present in sciatica and prolapsed intervertebral disc (PID). Tight hamstring muscles often interfere with the test.

Lasegue's sign—extension of the leg at knee, after flexing the thigh, will induce pain in PID.

Features of increased intracranial tension (IIT) :

(A) SYMPTOMS :

1. Headache (intense deep-seated dull aching pain, worsened by exertion or change of posture; headache may be severe on early morning hours).
2. Vomiting (sometimes, projectile; absence of nausea; unrelated to food).
3. Convulsions (usually generalised; focal due to SOL itself).
4. Altered consciousness to coma (retarded cerebration).
5. Drowsiness and mental deterioration may be present; visual problems like deterioration of visual acuity and peripheral constriction of visual field.

(B) SIGNS :

1. Pulse—Bradycardia; if the intracranial pressure continues to increase, the pulse becomes very rapid; there may be irregular pulse.
2. BP—There may be hypertension.
3. Respiration—Slow and deep respiration; later on it becomes rapid and shallow, and lastly Cheyne-Stokes breathing appears.
4. Papilloedema; pupil—unequal or abnormally reacting, may be fixed pupil.
5. False localising signs (signs which do not have localising value)—
 - (i) Unilateral or bilateral, VIth or IIIrd nerve palsy.
 - (ii) Bilateral Babinski's sign.
 - (iii) Bilateral grasp reflex.
6. Bulged anterior fontanelle in infants.
7. A 'chronic' IIT may produce features of 'hypopituitarism' e.g., loss of body hairs, hypothyroidism, hypoadrenalism etc.

* Examine—Pulse, pressure, plantar response, papilloedema and paralysis, if any (5P).

** Bradycardia, hypertension and irregular respiration are collectively known as "Cushing's triad" (due to cerebral herniation).

(C) CAUSES :

1. Cerebrovascular accidents (CVA).
2. Intracranial tumour.
3. Meningitis, meningism, encephalitis, cerebral abscess.
4. Benign intracranial hypertension (commonly from Addison's disease, corticosteroid withdrawal, COPD, overdose of vitamin A, hypoparathyroidism).
5. Circulatory block like aqueduct stenosis, 4th ventricle block in hydrocephalus.
6. Miscellaneous—Head injury, cerebral oedema, hypoxic encephalopathy etc.

(D) INVESTIGATIONS :

1. X-ray of skull —
 - (i) Silver-beaten appearance (in children),
 - (ii) Sutural diastasis (in children),
 - (iii) Erosion of clinoid process,
 - (iv) Deepened sella turcica,
 - (v) Enlargement of internal auditory meatus.
2. CT or MRI scan — Diagnoses IIT (dilatation of ventricles and effacement of cisterns) with the aetiology.

(E) TREATMENT :

It is done by propped-up position at 30°, I.V mannitol (20%), glycerol (oral or I.V), I.V steroid (dexamethasone in vasogenic oedema surrounding a tumour or abscess, but not in CVA), frusemide (I.V), retention enema of magnesium sulphate, or hyperventilation.

* As a false localising sign, VIth cranial nerve is commonly involved because it has the longest intracranial course and thereby easily compressed at the bony prominences. Lumbar puncture is contraindicated in IIT with papilloedema because of the risk of development of 'cerebellar pressure cone syndrome' (sudden death due to compression of vital medullary centres as a result of descent of cerebellar tonsil).

What are the common causes of coma ?

1. CVA (specially cerebral haemorrhage and massive cerebral infarction).
2. Encephalitis, meningitis, cerebral malaria, epilepsy, head injury, subdural or extradural haematoma, intracranial SOL, tentorial herniation.
3. Hypoglycaemia. diabetic ketoacidosis, renal failure, hepato-cellular failure, myxoedema coma, pituitary apoplexy, adrenal crisis.
4. Stokes-Adams syndrome, tight aortic stenosis, arrhythmias, respiratory failure (Type II).
5. Alcohol overdose, drug overdose, poisoning (barbiturate, opiate, organophosphorus), snake bite, hypothermia, heat stroke.
6. Psychogenic.

How do you like to investigate a case of hemiplegia ?

1. Blood (routine examination)—TC, DC, ESR.
2. Blood biochemistry—Sugar, lipid profile (cholesterol, LDL, VLDL, HDL, triglyceride), urea and creatinine.
3. ECG to diagnose myocardial ischaemia, chamber enlargement in heart, arrhythmia (atrial fibrillation) etc.
4. Chest X-ray (cardiac chamber enlargement, to diagnose pneumonia as a complication).
5. Echocardiography (to identify the source of emboli).
6. Carotid angiography and CSF study are not routinely done (carotid angiography may precipitate thrombosis in another territory of brain). Magnetic resonance (MR) angiography, digital subtraction angiography and lumbar puncture are diagnostic in subarachnoid haemorrhage.
7. Doppler flow studies in carotids (if facilities are available).
8. **CT scan of the brain**—For proper initial evaluation, CT scan is mandatory to categorise the stroke into either ischaemic (thrombosis or embolism) or haemorrhagic origin. Cerebral haemorrhages are detected immediately whereas infarction may take 48 hours or more. CT scan helps to decide the line of management (e.g., aspirin, anticoagulants or antiplatelet drugs). It also helps to diagnose the conditions which may simulate stroke e.g., ICSOL, vascular malformation etc. MRI scan may be done, though CT scan is preferred to MRI in acute stage.

* Read the special investigations from 'common causes of strokes in young' mentioned earlier.

What is the ideal line of management in hemiplegia ?

After initial evaluation of neurological deficit, cardiovascular status, metabolic status (blood sugar, urea, creatinine and electrolytes) and haematological parameters (Hb%, coagulation profile) one may start management of hemiplegia with general supportive measures, anti-oedema management, and care of the co-morbid conditions.

1. Patient should lie in a railed cot; insert Ryle's tube, put a self-retaining catheter and take care of the airway. Nursing cares should be started by trained persons. O₂ therapy, if necessary.
2. Treat **cerebral oedema** by,
 - a) 20 g mannitol 100 ml I.V thrice daily (reduces cerebral oedema by osmotic diuresis).
 - b) Oral glycerol—6 tsf thrice daily (usually taken with fruit juice).
 - c) Inj. dexamethasone (if not hypertensive) — 4-6 mg, I.M or I.V, every 4-6 hourly may be tried in selected cases e.g., chronic subdural haematoma (*contraindicated in CVA*).]
 - d) Inj. frusemide—40 mg I.V stat. and as and when necessary.
3. Treatment of diabetes mellitus, stress ulcer (by H₂RA or PPI), hypertension (maintain the BP at or just below 180 / 115 mm of Hg in cerebral infarction; and in cerebral haemorrhage, keep the BP a bit lower than the above reading), hyperlipidaemia; avoid smoking; maintain fluid balance. Do not lower the BP much in acute stage because it will hamper the cerebral autoregulation, diminish the perfusion of ischaemic zones, and may exacerbate infarction.
4. Antiplatelet drugs (in thrombosis)—Aspirin (75-300 mg daily) or dipyridamole (50 mg thrice daily) or ticlopidine (250 mg twice daily) for long term prophylaxis. Aspirin 300 mg daily should be started immediately as soon as the ischaemic stroke is confirmed, and reducing the dose to 75 mg daily after several days. Pentoxifylline (haemorrheology modifier, i.e., changes viscosity

of blood) may be beneficial in cerebral infarction if used within 12 hours of onset of stroke (not accepted by all). Newer antiplatelet drugs are clopidogrel (75 mg daily), lamifiban and abciximab. Aspirin is contraindicated in cerebral haemorrhage and very large infarction.

5. Role of cerebral vasodilators are contradictory (5% CO₂ with 95% O₂, cyclandelate, nicotinic acid, papaverine, aminophylline etc.) due to steal phenomenon.
6. Treatment of atrial fibrillation (in embolism) and valvular heart disease is done accordingly.
7. Anticoagulant therapy may be given (heparin and warfarin) in an 'evolving stroke' if cerebral haemorrhage is ruled out by early CT scan. Anticoagulant therapy is not of value in the treatment of completed stroke. Anticoagulation is mandatory in cardiogenic embolic stroke.
8. Recombinant tissue plasminogen activator (rTPA) may be beneficial if started within the first 3 hours of onset of an ischaemic stroke.
9. Surgery—Certain intracerebral haematomas are amenable to surgery (e.g., cerebellum); carotid endarterectomy is performed in carotid TIA.
10. Physiotherapy—Is done by an expert physiotherapist or physiatrist. **It should be started as soon as the patient recovers from the neural shock stage.** Change of posture in bed must be done to avoid bed sores. Physiotherapy is advised to prevent joint contractures and oedema of the limb, and to promote recovery of strength. Compression stockings should be applied to paralysed leg to prevent deep vein thrombosis. Passive movement of the limb also prevents venous stasis and pulmonary embolism. Chest physiotherapy should be started to promote coughing out of expectorations.
11. Rehabilitation; speech and occupational therapy should be tried.

* Avoid hypoxia, 5% dextrose infusion and corticosteroid in the treatment of CVA.

** Always take care of bladder, bowel, back (bed sore), base of the lungs, and boost the morale.

Case 17

Paraplegia

What is your diagnosis ?

This is a case of spastic paraparesis (or paraplegia) in extension due to compressive myelopathy caused by caries spine and the lesion is at the level of T₁₀ segment of the spinal cord.

Why do you say so ?

This young male patient aged 28 years was complaining of severe weakness of both the lower limbs with loss of sensation for last one month. He also complained of anorexia, cough with evening rise of temperature for last five months. One month back, he noticed gradual weakness of the left leg which was followed by weakness of the right leg after 2 days. He also noticed numbness or loss of superficial sensations like touch, pain and temperature below the level of umbilicus for the same duration. The fever was intermittent in nature and associated with night sweats. On repeated enquiry, he gave H/O **root pain**, i.e., sharp, constant, stabbing, electric shock-like pain which was distributed in the region below the umbilicus and both the lower limbs. The pain used to aggravate by the change of posture, coughing, sneezing, jolting and movement of spine. He also complained of a special type of sensation, i.e., girdle-like sensation at the level of umbilicus. For the last 25 days, there was almost total and more or less, symmetrical paralysis of both the lower limbs; there was no H/O diurnal variation of weakness (vide myasthenia gravis). The patient did not complain of any involvement or weakness of the upper limbs. It seems that the disease started insidiously and was progressing gradually.

Initially, he was also suffering from hesitancy of micturition and was catheterised in the hospital on admission (he was admitted in the hospital 25 days back). He was unable to hold faeces for the same duration (rectal incontinence). He specifically complained of bone pain at his back. Since his admission, there was no sign of recovery. The patient did not give any H/O flexor spasm.

There was no H/O headache, vomiting, convulsions, speech difficulty, facial weakness or any abnormal movements. No family H/O diabetes mellitus, hypertension or tuberculosis was found. There was no positive H/O exposure to STD, trauma to the spine or similar episodes in the past. He was not vaccinated in the recent past. He is neither diabetic nor hypertensive. On repeated enquiry, he gave H/O haemoptysis 3 months back which he ignored and did not attend any physician at that time.

On examination, it shows that the patient is conscious and co-operative. He is suffering from UMN paralysis of both the lower limbs. There is loss of superficial sensations below the umbilicus. A gibbus is

seen at the level of 7th thoracic vertebra which is tender on palpation. The UMN paralysis of both the lower limbs is suggested by bilateral clasp-knife spasticity, brisk tendon reflexes, presence of clonus and extensor plantar response.

* Paraplegia is basically of two types : Spastic and flaccid.

What is your case ?

Say the summary as above.

Importance of 'past history' in paraplegia :

1. H/O tuberculosis.
2. H/O fever (tuberculosis, acute transverse myelitis, G. B. syndrome, lymphoma, leukaemia).
3. Primary chancre (exposure to STD).
4. Similar episodes in the past (may signify multiple sclerosis).
5. Spinal injury (produces compressive myelopathy).
6. Hypertension, diabetes mellitus as a routine.
7. H/O any lymphadenopathy (tuberculosis, lymphoma, malignancy) or vaccination (may precipitate acute transverse myelitis-like features).
9. Alcoholism or drugs like INH, vincristine (peripheral neuropathy producing weakness of both lower limbs).
10. Pain in the spine (caries spine, metastasis in the spine).
11. Occupational history : H/O exposure to chemicals and toxins

Importance of family history' in paraplegia :

1. Hypertension (as a routine).
2. Diabetes mellitus (as a routine).
3. H/O paraplegia in other members of the family (if present, may indicate hereditary spastic paraplegia, paraplegia with hereditary ataxia, or lathyrism).
4. Tuberculosis.

Chief complaints (clinical presentations) of your patient :

1. Weakness of both the lower limbs for 1 month.
2. Loss of sensations of both the lower limbs for 1 month.
3. Hesitancy of micturition and loss of bowel control for 1 month.
4. Cough with evening rise of temperature for last 5 months.

Enquiry regarding paralysis (in H/O present illness) :

(A) *DATE OF ONSET* : Note the specific date and time of onset of paralysis.

(B) *MODE OF ONSET* : Sudden (acute transverse myelitis, spinal injury etc) or gradual (caries spine).

(C) *PRECIPITATING FACTORS* :

- (i) Spinal injury.
- (ii) Vaccination (commonly anti-rabies nervous tissue vaccine).

(D) *EVOLUTION OF PARALYSIS* : Whether both the limbs are affected simultaneously or one after the other.

(E) *PROGRESS OF PARALYSIS* :

- (i) Increasing in severity and extent—Cord compression.
- (ii) Improving—Inflammatory, acute transverse myelitis or multiple sclerosis.
- (iii) Static but progressing very slowly—Degenerative lesion (Friedreich's ataxia).
- (iv) Waxing and waning—Multiple sclerosis.

(F) *DEGREE AND DURATION OF PARALYSIS*—Weakness or total loss of power, with duration.

(G) *MOTOR SYMPTOMS*—See the symptomatology.

Enquiry regarding sensory symptoms (in H/O present illness) :

1. Loss of sensation (superficial and/or deep)—find out the upper border.
2. Girdle-like sensation or sense of constriction (find out the level).
3. Zone of hyperaesthesia (find out the level).
4. Root pain (ask for distribution and precipitating factors).
5. Sensation of pins and needles in the lower extremities (test for sensory functions).

Cerebral paraplegia :

(A) Causes : The lesion in paraplegia lies either in cerebral cortex, spinal cord, nerve roots, peripheral nerves, myoneural junction, or in muscles. Cerebral causes are extremely rare and always give rise to paraplegia in extension. The causes are,

1. Cerebral diplegia (Little's disease) — Cortical cause of paraplegia.
2. Superior sagittal sinus thrombosis.
3. Parasagittal meningioma.
4. Thrombosis of unpaired anterior cerebral artery.
5. Hydrocephalus.
6. Bullet injury at paracentral lobule.

(B) Features :

1. Extremely rare. H/O jacksonian fits and cortical sensory loss present.
2. Spastic paraplegia in extension (may be flaccid in shock stage but never produces paraplegia in flexion).
3. Features of IIT may be present (headache, vomiting, convulsions).
4. Abnormalities in higher function, speech difficulty, cranial nerve affection may be present.

Table 9 : Differentiation between 'paraplegia in extension' and 'paraplegia inflexion'

Spastic paraplegia is of two types : Paraplegia in extension and paraplegia in flexion.

Features	Paraplegia in extension	Paraplegia in flexion
1. Definition—	1. Lower limbs take an extension attitude and the extensor muscles are spastic	1. Lower limbs take an attitude of flexion due to ' flexor spasm ' (even the heel may strike the back of ipsilateral thigh)
2. Pathology—	2. Only pyramidal tracts are involved	2. Both pyramidal and extrapyramidal tracts (e.g., reticulospinal tract) are involved (thus, prognostically worse). Occurs in late stages of paraplegia or progressive lesion
3. Evolution—	3. Early	3. Late
4. Clinical features—		
a) Attitude	a) Hip extended and adducted, knee extended, and feet plantiflexed	a) Thigh and knee flexed, feet dorsiflexed. Adopts lateral decubitus posture in bed
b) Tone	b) Clasp-knife spasticity in extensor group of muscles	b) Tone is increased in flexor group of muscles
c) Jerks	c) Brisk	c) Present but diminished
e) Plantar response	e) Extensor	e) Extensor but may be associated with flexor spasms
f) Reflex evacuation of bladder and bowel	f) Absent	f) Present
g) Mass reflex	g) Absent	g) May be present

**** Mass reflex :** If the skin of the lower limbs or the lower abdominal wall is stimulated, there is reflex flexion of the lower trunk muscles and the lower limbs (ie, flexor spasm), piloerection and penile erection, evacuation of the bladder, bowel and semen, and sweating. Mass reflex indicates severe spinal cord lesion. It is a reflex of spinal automatism.

*** Flexor spasm may be severe in posterior column lesion (e.g., multiple sclerosis), in the presence of bed sores or urinary tract infection (as a result of constant stimulation of small unmyelinated nerve fibres) with cord lesion.

Causes of 'spastic' paraplegia (UMN type lesion) :

(A) GRADUAL ONSET :

- I. Cerebral causes—Cerebral diplegia, parasagittal meningioma, hydrocephalus.
- II. Spinal causes—
 - a) Compressive or transverse lesion in the spinal cord—Cord compression (see below).
 - b) Non-compressive or longitudinal lesion or systemic diseases of the spinal cord
 - (i) Motor neurone disease (MND) e.g., amyotrophic lateral sclerosis.
 - (ii) Multiple sclerosis, Devic's disease.
 - (iii) Friedreich's ataxia.
 - (iv) Subacute combined degeneration (i.e., from vitamin B₁₂ deficiency).
 - (v) Lathyrism.
 - (vi) Syringomyelia.
 - (vii) Hereditary spastic paraplegia.
 - (viii) Syphilitic meningomyelitis (rare).
 - (ix) Tropical spastic paraplegia.
 - (x) Paraneoplastic myelitis.
 - (xi) Antiphospholipid antibody syndrome.
 - (xii) Radiation myelopathy.

(B) SUDDEN ONSET:

- I. Cerebral causes—Thrombosis of unpaired anterior cerebral artery, superior sagittal sinus thrombosis, bullet injury at paracentral lobule.
- II. Spinal causes—
 - a) Acute transverse myelitis.
 - b) Injury to the spinal cord (fracture-dislocation or collapse of the vertebra).
 - c) Thrombosis of anterior spinal artery (myelomalacia).
 - d) Haematomyelia (from arteriovenous malformations, angiomas, or endarteritis).
 - e) Post-vaccinal.
 - f) Prolapsed intervertebral disc (PID).
 - g) Spinal epidural abscess or haematoma.
 - h) Radiation myelopathy.

* Compressive—II b), f) and g); Non-compressive—II a), c), d), e) and h).

Causes of 'cord compression' (compressive aetiology in paraplegia) :

I. Intramedullary (5%) :

- a) Glioma.
- b) Ependymoma.
- c) Chordoma.
- d) Syringomyelia, haematomyelia.

n. Extramedullary :

- a) **Intradural (15%)—**
 - (i) Meningioma.
 - (ii) Neurofibroma.
 - (iii) Patchy arachnoiditis (tuberculosis, syphilis, sarcoidosis).
 - (iv) Arteriovenous malformations.
- b) **Extradural (80%)—**
 - (i) Caries spine (Pott's paraplegia).
 - (ii) Myeloma, lymphomatous or metastatic deposits in the vertebra; osteomyelitis.
 - (iii) Patchy meningitis.
 - (iv) Prolapsed intervertebral disc (PID).

- (v) Degenerative joint disease (spondylosis).
- (vi) Fracture or dislocation of the vertebra, osteoporosis, paget's disease.
- (vii) Extramedullary haematopoiesis in thalassaemia.
- (viii) Spinal epidural abscess or haematoma.

* Cord compression produces paraplegia by : 1. Pressure effect, 2. Ischaemia (arterial), and 3. Congestion (venous).

** Intradural lesion (e.g., neurofibroma) gives a history of long duration while extradural lesion (e.g., malignancy) gives a short history.

Causes of flaccid' paraplegia (LMN type lesion) :

(A) *UMN LESION IN SHOCK STAGE*—i.e., sudden onset spastic paraplegia in 'neural shock' stage e.g., acute transverse myelitis, spinal injury.

(B) *LESION INVOLVING ANTERIOR HORN CELLS* —

- a) Acute anterior poliomyelitis.
- b) Progressive muscular atrophy (variety of MND).
- c) Trauma.

(C) *DISEASES AFFECTING NERVE ROOTS*—Tabes dorsalis, radiculitis, G.B. syndrome.

(D) *DISEASES AFFECTING PERIPHERAL NERVES*—

- a) Acute infective polyneuropathy (G.B. syndrome).
- b) High cauda equina syndrome.
- c) Disease of peripheral nerves involving both the lower limbs.
- d) Lumbar plexus injury (e.g., posas abscess or haematoma).

(E) *DISEASES AFFECTING MYONEURAL JUNCTION*—

- a) Myasthenia gravis, Lambert-Eaton syndrome.
- b) Periodic paralysis due to hypo- or hyperkalaemia.

(E) *DISEASES AFFECTING MUSCLES*—Myopathy.

(F) *HYSTERICAL PARALYSIS*.

* Note that paraplegia may be produced due to lesion in cerebral cortex, spinal cord (compressive or non-compressive myelopathy), nerve roots, peripheral nerves, myoneural junction or muscles. In clinical practice, we are more concerned with paraplegia due to lesions in the spinal cord (myelopathy).

* *Common causes of paraplegia are 5T: Trauma, tuberculosis, tumour, thrombosis and transverse myelitis.*

Classical features of acute transverse myelitis :

Though a common cause of non-compressive myelopathy (acute onset total transection of the cord), clinically somewhat **it behaves like compressive variety**.

1. Very often follows a viral illness (neurotropic virus) or post-vaccinal.
2. Onset is acute or subacute (evolve over several days to 2-3 weeks).
3. **Fever may be present before paralysis develops.**
4. Bladder involvement is early.
5. Though behaves like compressive variety, usually there is absence of root pain, spinal tenderness or spinal deformity. Back pain and progressive paraparesis are presenting complaints. Girdle constriction at the level of lesion with zone of hyperaesthesia just above may be obtained. Mid-thoracic region is the most common site.
6. Plantar response is extensor and there is partial or complete sensory loss (all modalities), with a definite upper level (very important **point of difference with acute infective polyneuropathy** where these two features are, flexor and no demonstrable sensory loss respectively).
7. MRI scan is often required to exclude cord compression.
8. Majority (70%) recovers within 12 weeks.

* Plantar response may not be extensor, in the stage of neural shock.

** All the reflexes are usually lost below the level of lesion in the stage of neural shock (first 2-3 weeks). Thereafter, spasticity with brisk reflexes develop.

*** Other causes of acute transverse myelopathy are multiple sclerosis, vasculitis (MCTD), HIV, sarcoidosis, radiation, syphilis and anterior spinal artery occlusion.

Features of acute infective polyneuropathy (Landry-Guillain-Barre syndrome) :

1. There is an association with Epstein-Barr virus, measles, campylobacter jejuni (causing diarrhoea), HIV, cytomegalovirus, post-vaccinal and post-surgical events. The current hypothesis is in the favour of immunological reaction due to hypersensitivity, perhaps as a result of unidentified allergen directed against the myelin sheath of peripheral nerves. Truly speaking, it is an acute post-infective **polyradiculoneuropathy** (i.e., inflammatory demyelination involves spinal roots and peripheral nerves). Severe forms show secondary axonal degeneration.
2. a) Common history :
Viral illness (e.g., respiratory catarrh) -> after 2 weeks, paralysis with sensory symptoms (paraesthesia) starts -> paralysis progresses for 3 weeks and then stops -> paralysis becomes static for 2 weeks -> after 2 weeks recovery starts which is usually completed within 6 months -> in incomplete recovery, residual disability persists,
b) Incidence — in either sex; peak incidence is in between 20-50 years.
3. Monoplegia, paraplegia or quadriplegia (**proximal muscles are more affected**). It is an acute (evolved over hours to days) **ascending LMN type of paralysis usually beginning in the legs**. Upper extremities may also be involved leading to quadriplegia. There may be bulbar palsy or respiratory muscle paralysis.
4. Bilateral VIIth (LMN type) cranial nerve palsy (present in approximately 50% cases), though this is occasionally unilateral.
5. No sensory abnormalities demonstrated (very characteristic) though sometimes the patients complain of distal paraesthesia.
6. Jerks are invariably absent (i.e., there is diffuse weakness with widespread loss of reflexes). Gross hypotonia is evident.
7. Bladder and bowel are rarely involved.
8. Plantar response—Flexor or no response.
9. Autonomic disturbances (tachycardia, arrhythmia, postural hypotension/hypertension) are not uncommon.
10. Sensorium (consciousness)—Clear throughout the disease process.

* **There is subjective sensory symptoms (paraesthesia) without any objective sensory loss.**

** G.B. syndrome is also known as acute inflammatory demyelinating polyneuropathy (AIDP).

*** Outcome : 1. Complete or near complete recovery - 80%, 2. Residual disability - 15%, and 3. Death - 5% (respiratory paralysis or lung infection).

**** It is an areflexic, atonic ascending motor paralysis.

Features of 'atypical' G.B. syndrome :

- | | |
|---|---|
| 1. Distal muscle involvement. | 7. Glomerulonephritis. |
| 2. Objective sensory impairment. | 8. Miller Fisher syndrome (ataxia, areflexia and external ophthalmoplegia) without significant limb weakness. |
| 3. Papilloedema. | 9. SIADH (syndrome of inappropriate ADH secretion). |
| 4. Descending paralysis. | |
| 5. Early bladder and bowel involvement. | |
| 6. Babinski's sign. | |

What is Brown-Sequard syndrome ?

It is the hemisection of the spinal cord (e.g., bullet or stab injury) and the features are :

1. Sensory—Loss of proprioception (joint, position, vibration sense) on the same side, and loss of pain and temperature sensations on the opposite side, 1-2 segment below the level of lesion.
2. Motor—Segmental LMN signs at the level of lesion; monoplegia of the lower limb or hemiplegia on the same side may occur below the level of lesion.
3. Reflexes—Below the level of lesion, there are features of UMN lesion, i.e., superficial reflexes lost, brisk deep reflexes with extensor plantar response.

* There may be a band of hyperaesthesia on the same side just above the level of lesion.

** In lesion above T₁ level ; patient presents with Horner's syndrome.

*** **Maximum** motor loss on the same side and maximum sensory loss on the opposite side.

Localisation of the level of lesion in a compressive myelopathy :

Localisation of the level of compression is done by :

1. Distribution of root pain—Ask for specific dermatomes involved (it is due to involvement of posterior nerve roots).

2. Upper border of sensory loss—Examine the patient from below upwards (i.e., from legs to thorax) for demonstration of upper border of sensory loss (due to affection of spinothalamic tract).
 3. Girdle-like sensation or sense of constriction at the level of lesion (usually due to involvement of posterior column).
 4. Zone of hyperaesthesia or hyperalgesia—This will localise the level of lesion one segment below i.e., zone of hyperaesthesia is present just above the level of girdle-like sensation, and is due to compression of posterior nerve roots.
 5. Analysis of abdominal reflex—i.e., if upper abdominal reflex is intact with loss of middle and lower one, the site of lesion is probably at T₁₀ spinal segment.
 6. Atrophy of the muscles in a segmental distribution (due to involvement of anterior horn cells).
 7. Loss of deep reflexes, if the particular segment is involved. The reflexes will be brisk below the involved segment.
 8. Analysis of *Beevor's sign* (do the 'rising test'. In paralysis of lower part of rectus abdominis, umbilicus moves upwards and in paralysis of upper part of rectus, umbilicus goes downwards)—Beevor's sign detects the weakness of abdominal muscles.
 9. Deformity or any swelling in the vertebra.
 10. Tenderness in the vertebra.
 11. The area of sweating may help (lack of sweating below the level) in localising the level of lesion.
 12. The level can also be localised by X-ray of the spine, myelography, CT scan or MRI.
- * The question of localisation of the level of lesion does not arise in non-compressive myelopathy.

Determination of spinal segments in relation to vertebra :

The spinal segments do not lie exactly under the overlying vertebra. The following formula determines the spinal segments related to a vertebral body.

1. For cervical vertebrae—add 1.
2. For 1-6 thoracic vertebrae—add 2.
3. For 7-9 thoracic vertebrae—add 3.
4. The T₁₀ arch overlies L₁ and L₂ segments.
5. The T₁₁ arch overlies L₃ and L₄ segments.
6. The T₁₂ arch overlies L₅ segment.
7. The L₁ arch overlies the sacral and coccygeal segments.

N.B.: In the patient described at the outset, the T₇ vertebra is involved and thus, the spinal segment involved is T₁₀ (by adding 3). Remember, in lower thoracic region the tip of a spinous process marks the vertebral body below.

* Altogether 31 pairs of spinal nerve roots emerge from the spinal cord. The segments in the cord are : cervical 8, thoracic 12, lumbar 5, sacral 5 and coccygeal 1. The lower end of spinal cord tapers to form 'conus medullaris' at lower border of L₁ vertebra. The spinal roots below L₁ vertebra pass down in the spinal canal to emerge at their respective foramina, and thus form 'cauda equina'.

Points to note in spine examination in paraplegia :

- I.
 1. Kyphosis.
 2. Scoliosis.
 3. **Gibbus** (with paraspinal muscle spasm).
 4. Tenderness.
 5. Spina bifida.
- II. Any scar (indicates old trauma), tuft of hair over sacrum (spina bifida occulta), bed sore or sacral oedema.

'Paraplegia in extension' progressing to 'paraplegia inflexion' : How to diagnose?

1. Appearance of flexor spasms.
2. Diminution of deep reflexes (jerks).
3. Diminution of ankle clonus in duration.

* In addition to pyramidal tract, there is involvement of extrapyramidal tracts like reticulospinal, rubrospinal, tectospinal or vestibulospinal tract in patients with paraplegia in flexion.

** Pathophysiology : when the pyramidal tract alone is at jeopardy, the extrapyramidal system takes

the upper hand, resulting in increased tone of the antigravity muscles (i.e., paraplegia in extension). When the extrapyramidal system is at fault too, the spinal arc becomes dominant and there is relative increase in tone of the flexors (e.g., hamstrings) more than the extensors (i.e., paraplegia in flexion).

Table 10 : Differentiation between compressive and non-compressive myelopathy

Features	Compressive	Non-compressive
1. Bony deformity	+	-
2. Bony tenderness	+	-
3. Girdle-like sensation	+	-
4. Upper level of sensory loss	+	-
5. Zone of hyperaesthesia	+	-
6. Root pain	+	-
7. Onset and progress	Gradual	May be acute
8. Symmetry	Asymmetrical	Majority are symmetrical
9. Flexor spasm	Common	Usually absent
10. Pattern of neurodeficit	** U-shaped	Bilaterally symmetrical
11. Bladder and bowel involvement	Early	Late (early in acute transverse myelitis)
12. Classical example	Caries spine	Motor neurone disease

* No. 3, 4 and 5 are sometimes present in acute transverse myelitis.

** March of neurodeficit pattern : U-shaped, i.e., first ipsilateral upper limb (UL), next ipsilateral lower limb (LL), next contralateral LL, and finally contralateral UL (Elsberg's phenomenon). In a slow cord compression the pyramidal tract is affected first, followed by the posterior column, and finally the spinothalamic tract.

Table 11 : Discriminate between extramedullary (intradural) and intramedullary lesions

Features	Extramedullary	Intramedullary
1. Root pain	Early and common	Root pain is rare; pain is burning in type and poorly localised
2. Sensory deficit	No dissociation of sensation; contralateral loss of pain and temperature with ipsilateral loss of proprioception	Dissociation of sensation common; spotty changes
3. Sacral sensation	Lost (early)	Sacral sparing
4. LMN involvement	Segmental	Marked with widespread atrophy; fasciculation seen
5. UMN involvement	Early and prominent	Less pronounced; late feature
6. Reflexes	Brisk, early feature	Less brisk, late feature
7. Autonomic involvement (bladder and bowel)	Late	Early
8. Trophic changes	Usually not marked	Common
9. Vertebral tenderness	May be sensitive to local pressure	No bony tenderness in vertebrae
10. Changes in CSF (i.e., raised proteins)	Frequent	Rare

* Sacral sensation : in extramedullary lesion (e.g., cauda equina lesion) lower sacral sensations are impaired (i.e., saddle anaesthesia), and in intramedullary lesion sensation is preserved in sacral dermatomes (sacral sparing).

Table 12 : Differentiation between cauda equina syndrome and conus medullaris lesion

Features	Cauda equina syndrome	Conus medullaris lesion
1. Onset	Gradual and unilateral (or asymmetrical involvement of both lower limbs)	Sudden and bilateral
2. Aetiology	PID, metastases	Intramedullary SOL (e.g., glioma)
3. Root pain	Severe low backache	Usually absent
4. Motor involvement	Asymmetric limb weakness	Muscle changes are not marked
5. Sensory loss	Asymmetric sensory loss (i.e., unilateral saddle anaesthesia)	Bilateral saddle anaesthesia
6. Bladder and bowel function	Late and less marked	Early and marked
7. Anal reflex (S ₃₋₄)	Areflexia if roots are involved	Anal reflex absent
8. Deep reflexes	Ankle and knee jerks are lost	Ankle and knee jerks are usually preserved
9. Trophic changes	Less	Early
10. Plantar response	Flexor or no response	Extensor (not always)

* Cauda equina lesion usually gives rise to LMN signs whereas conus medullaris lesion have UMN signs.

Abdominal reflex and its interpretations :

It is a polysynaptic nociceptive reflex. The clinical method goes like this :

1. The patient lies supine with exposed abdomen. The procedure is explained to him. Now the **stroke is given bilaterally in 3 places**—below the costal margins, at the level of the umbilicus and above the inguinal ligaments. Always compare right with the left side. Stroke is done lightly by a needle, a key or by the shaft of the hammer.
2. Stroke is given from outside to the centre on both the sides sequentially (the dermatomes are criss-crossed at the midline and if the stroke starts from midline, eventually by chance, the opposite side may be stimulated). The hammer stick should be placed at an acute angle with the abdominal skin and it should not cause any abrasion in the skin.
3. The normal reflex involves contraction of anterior abdominal musculature with movement of umbilicus in that direction. The reflex can be read by observing the movement of umbilicus (elevation, depression or lateral deviation) which is known as Beevor's sign.

Root value : T₈ - T₁₂.

Loss of abdominal reflex :

(a) Physiological :

- (i) Anxiety.
- (ii) Obesity.
- (iii) Lax abdominal wall.
- (iv) Multiparous woman.
- (v) Aged patient.
- (vi) In the presence of ascites or pregnancy.
- (vii) Previous abdominal surgery.

(b) Pathological :

- (i) LMN type of lesion affecting the corresponding reflex arc.
- (ii) Peripheral neuropathy.
- (iii) UMN type of lesion above the level of the reflex arc (hemiplegia or myelopathy).

* Abdominal reflex is lost early in multiple sclerosis, and retained long in MND and cerebral diplegia. In hemiplegia, the abdominal reflex is lost in the paralysed side only. Over-response occurs in psychoneurosis. Abdominal reflex (superficial reflex) is a local reflex arc modified by a 'cerebral arc' i.e., the pyramidal tract.

How cremasteric reflex is elicited ?

Scratch the medial side of upper thigh in a downward and inward direction (thigh remains abducted and externally rotated), and the response is upward movement of ipsilateral scrotum and testicle due to contraction of cremasteric muscle. Alternatively, this reflex can be easily elicited by pressing over the sartorius in the lower third of Hunter's canal. The spinal segment responsible are **L₁** and **L₂**. This reflex

is lost in either UMN lesion or damage to L₂ spinal segments, or with hydrocele/hernia. It is also a polysynaptic reflex. Often it is very difficult to elicit this reflex in the elderly.

* Different superficial reflexes are : corneal and conjunctival reflex, abdominal reflex, cremasteric, and plantar reflex.

Special points in the physical examination in paraplegia :

(A) GENERAL SURVEY :

- a) The patient is conscious and co-operative.
- b) Decubitus—Lying helplessly without any movement of the lower limbs and with extension of all joints (lower limb). Upper limbs are within normal limit.
- c) Lymph nodes—Not palpable (ref : tuberculosis, lymphoma, malignancy or HIV infection).
- d) Pulse—Mention all the points.
- e) BP—It is compulsory to record the BP (in all long cases); 125/85 mm of Hg in the present case.
- f) Respiration—18/min, regular, abdominal in type (ref. : respiratory depression in G. B. syndrome).
- g) Temperature—Record the oral temperature (tuberculosis, acute transverse myelitis, UTI etc.).
- h) Oedema—Not present (may be present due to prolonged recumbency). Look for sacral oedema.
- i) Skin—Absence of neurofibroma or cafe-au-lait spot.

* Mention all the points in general survey.

(B) NERVOUS SYSTEM :

- a) The patient is conscious, alert, oriented and co-operative.
- b) Higher functions : Mention all the subheadings—All are normal here; speech—Normal.
- c) Cranium and spine ; Cranium is normal and there is presence of gibbus (T₇) with spinal tenderness.

* **One must examine the spine in all patients with paraplegia.**

- d) Neck rigidity, Kernig's sign ; Absent; straight leg raising test—negative.
- e) Cranial nerves ; No paralysis detected. Always search for optic atrophy (e.g., Friedreich's ataxia, multiple sclerosis) and VIIth nerve palsy (G.B. syndrome). Leukaemic or lymphomatous deposits may produce unilateral or bilateral VIIth nerve palsy (LMN type) with cord compression. Papilloedema may be found in G. B. syndrome. Mention the subheadings under the different cranial nerves.
- f) Motor functions :
Both the upper limbs are within normal limit in all respect.
 - (i) Nutrition—Helpless attitude of both the lower limbs; no atrophy at present.
 - (ii) Tone—Clasp-knife spasticity in both the lower limbs; extensor tone > flexor tone.
 - (iii) Power—Grade I in left leg and Grade II in right leg.
 - (iv) Coordination—Upper limbs ; Normal.
Lower limbs : Could not be tested,
- v) Involuntary movements—None present (search for fasciculation in the upper and lower limbs—MND). No flexor spasm seen.
- g) Sensory functions :
Both the upper limbs are normal.

<ol style="list-style-type: none"> (i) Superficial—Pain, touch and temperature (ii) Deep—Joint sense, position sense, muscle sense, pressure sense and vibration sense (iii) Cortical sensation—Point localisation, stereognosis 	}	All are lost in both the lower limbs
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* **There is a definite line of demarcation of sensory loss at the level of umbilicus (T₁₀). Below the level of umbilicus, there is no sensation present.**

h) Reflexes :

(i) Superficial—

- a) Abdominal—
 - x) Upper—Present bilaterally,
 - y) Middle—Lost bilaterally,
 - z) Lower—Lost bilaterally.
- b) Cremasteric—lost bilaterally.
- c) Plantar response—**Extensor in both the lower limbs.**

Deep—	Right	Left
a) Biceps jerk	Normal	Normal
b) Triceps jerk	Normal	Normal
c) Supinator jerk	Normal	Normal
d) knee jerk	Brisk	Brisk
e) Ankle jerk	Brisk	Brisk
f) Clonus (ankle and patellar)	Present	Present

* Mention the jaw jerk under Vth cranial nerve.

(iii) Visceral—

- a) Bladder—Catheterised at present. A self-retaining catheter with continuous drainage into a urosac is seen.
- b) Bowel—Incontinence at present.
- c) Swallowing—Normal at present.

(iv) Other reflexes—Glabellar tap, grasp reflex, palmo-mental reflex are WNL.

* Percuss the urinary bladder and confirm whether it is distended or not. Always see the colour of the urine present in the tube or urosac (chalky or turbid urine is seen in UTI, orange coloured urine is a clue to intake of rifampicin).

- i) Trophic changes : Bed sore present over sacrum (**one must search for the trophic changes in paraplegia**).
- j) Cerebellar functions : Normal (examinations involving lower limbs could not be performed),
- k) Autonomic functions : Loss of sweating below the level of umbilicus.
- l) Gait : Could not be tested.

* **Sacrum should be examined in paraplegia for the presence of oedema and/or bed sore.**

(C) *RESPIRATORY SYSTEM : SEARCH FOR—*

- (i) Tuberculosis—Crepitations at the apex, consolidation or pleural effusion.
- (ii) Bronchogenic carcinoma—Mass or collapse, pleural effusion, SVC syndrome.
- (iii) Lymphoma—SVC syndrome or pleural effusion.

* Format of examination of 'Respiratory system' is similar to 'Hemiplegia'.

(D) *G.I. TRACT AND GENITOURINARY SYSTEM* : Palpate liver, spleen and examine for ascites (carcinoma, tuberculosis, lymphoma). Percuss for bladder fullness.

(E) *CVS* : As in 'Hemiplegia'.

(F) *LYMPHORETICULAR SYSTEM* : Examine for sternal tenderness, haemorrhagic spots, lymphadenopathy etc.

N.B. : Special points in nervous system (may be added in history sheet) : Structures involved in this patient—corticospinal tract, lateral spinothalamic tract, posterior column and T₁₀ nerve roots (posterior).

Grading of muscle power in neurology (by Medical Research Council Scale) :

Grade 0 — Complete paralysis.

Grade 1 — A flicker of contraction only (visible or palpable) without any movement of joint.

Grade 2 — Movements possible only after elimination of gravity (side to side movement of a limb).

Grade 3 — Movements possible against gravity but not against resistance.

Grade 4 — Movements possible against gravity plus resistance but weaker than normal.

Grade 5 — Normal power.

* Often, '+' or '-' symbols are used to improve the sensitivity of the grading.

** Weakness of small muscles of hand and foot does not require grading.

How do you assess motor functions in neurology ?

Motor system is assessed by these 5 points :

1. Nutrition or bulk of muscles (i.e., inspection and palpation of muscles).
2. Tone of muscles.
3. Power of muscles.
4. Coordination of movement.
5. Involuntary movements.

* Examination of **reflexes** and **gait** are additional points in motor function.

Assessment of the nutrition of muscle :

Note the nutrition of muscles and observe the 'attitude' of the limb. It is assessed by :

(A) Inspection—

- (i) Wasting or atrophy of muscles (e.g., convexity of deltoid or calf muscle is lost in wasting).
- (ii) Flattening of overlying skin or hollowness over the area.
- (iii) Prominent knuckles and other bony prominences.
- (iv) Prominent interosseous gutters (in hand and foot).
- (v) Prominent extensor/flexor tendons (in hand or foot).

N.B. : Examine the big muscles like biceps, quadriceps, calves plus small muscles of the hands and feet. During inspection, other than '**attitude of a limb**', one should look for atrophy, hypertrophy, fasciculations, contracture, deformity or flexor spasm.

(B) Palpation—Atrophied muscles are small, soft and flabby on palpation. Normal muscle is elastic in feel.

(C) Measurement—Measure the girth of the specific muscle by a tape from a fixed bony point and compare it with the identical point on the other side (**compulsory**). For the upper limb, the fixed bony point is the elbow (olecranon process) and for the lower limb, it is the tibial tuberosity. The difference in the circumference (comparing with the opposite side) will give objective evidence of wasting or hypertrophy. Remember, in a right-handed person the girth of right upper limb may be slightly more than that of left upper limb.

* Muscles feel doughy (rubbery) in pseudohypertrophic muscular dystrophy, semi-elastic in true hypertrophy, tender in myositis, hard and inelastic in polymyositis, firm in spasticity, and 'woody' in rhabdomyolysis.

** If a patient have muscle atrophy in limbs, it is mandatory to measure the circumference of limbs in long or short cases.

Table 13 : Differentiation between muscle 'atrophy due to LMN lesion' and 'disuse atrophy'

Atrophy (LMN lesion)	Disuse atrophy
1. Usually atrophy occurs from distal to proximal muscles of a limb	1. Occurs in antigravity muscles of a limb (e.g., in hemiplegia)
2. Muscle power is very much reduced	2. Power loss is less than LMN lesion
3. Fasciculations may be evident	3. Fasciculations are never seen
4. Trophic changes may develop	4. Trophic changes are usually absent.
5. Deep reflexes or jerks are lost or diminished	5. Deep reflexes or jerk may be brisk

Causes of hypo- and hypertonia :

Tone—It is defined as the resistance felt when a joint is moved passively through its range of movement, or it is the partially contracted condition of a muscle. Tone is of three types : normal (elastic type of resistance), hypotonia and hypertonia. Muscle tone is basically regulated by pyramidal and extrapyramidal pathways.

(A) Hypotonia (diminished resistance to passive movements) :

1. LMN lesion.
2. Posterior column lesion (tabes dorsalis).
3. UMN lesion in shock stage.
4. Chorea (the only extrapyramidal lesion with hypotonia).
5. Cerebellar lesion.
6. Myopathy (terminal stage).
7. Myasthenic crisis.
8. Down's syndrome.
9. Rickets.
10. Deep sleep.
11. Drugs—Hypnotics, sedatives, muscle relaxants and anaesthetics.
12. Periodic paralysis, cataplexy, sleep paralysis.

13. Hypokalaemia or hypercalcaemia.

14. Emaciation.

* **LMN lesions** include anterior horn cell disease (poliomyelitis), anterior nerve root lesion or radiculopathy (arachnoiditis, radiculitis, G. B. syndrome), nerve trunks and plexus lesion (metastasis, trauma), disorders of peripheral nerves (leprosy), diseases of myoneural junction (myasthenia gravis) and muscle disease (late stage of myopathy). Hypotonia is also known as '**FLACCIDITY**'. Hypotonia is usually associated with hyporeflexia, muscle wasting and weakness.

** Affection of anterior horn cells is known as 'neuronopathy', and that of plexus (brachial/lumbosacral) is termed as 'plexopathy'.

(B) **Hypertonia** (increased resistance to passive movements) :

1. UMN lesion (clasp-knife spasticity).
2. Extrapyramidal lesion except chorea (lead pipe or cogwheel rigidity)—e.g., parkinsonism.
3. Tetanus.
4. Hysterical hypertonia (resistance to passive movement increases in an irregular, jerky fashion in proportion to the effort applied by the examiner).
5. Sometimes in non-cooperative patients.
6. Tetany.
7. Myotonia during the contraction phase.
8. Strychnine poisoning.
9. Gegenhalten phenomenon (variable resistance found in catatonic states e.g., encephalopathy, carbon monoxide poisoning, phenothiazine-induced; dementia)—classically a manifestation of frontal lobe disease where flexors and extensors are equally affected.
10. Decerebrate rigidity (there is marked contraction of all extensor muscles in the body).
11. Catatonia—a form of psychosis where the patient is hypomobile and mute. There is 'waxy flexibility' of the limbs.
12. Muscles around a painful joint.

*** In health, tone is being inhibited by pyramidal tract, and vestibulospinal and reticulospinal tracts; thus hypertonia occurs in UMN and extrapyramidal lesions.

**** Hypertonia is of two types—**SPASTICITY** (pyramidal lesion) and **RIGIDITY** (extrapyramidal lesion). Spasticity is usually associated with brisk tendon reflex, clonus, Babinski's sign and classical pattern of weakness. Rigidity (e.g., as seen in parkinsonism) is frequently associated with bradykinesia, static tremor and postural abnormality. 'Reflex rigidity' is the muscle spasm in response to pain e.g., neck rigidity in meningitis, board-like rigidity in peritonitis.

Features of hypotonia :

1. Unusual flexibility of joints leading to **increased** range of passive movement.
2. Inability to maintain sustained posture.
3. Muscles are soft and flabby, and smaller than normal.

* In **hypertonia** : Muscles feel stiff and there is **diminished** range of passive movement.

Clinical assessment of tone of muscles :

Tone may be **assessed at the bedside by the following methods :**

1. Classical method—By handling the limbs and moving them passively at their various joints (to feel the resistance to passive movement)—**best method**. While testing the 'tone', ask the patient to relax and be comfortable, and
 - (i) Expose the muscle fully.
 - (ii) Passively move the joint concerned.
 - (iii) Feel the resistance to stretching during the passive movement and see the contraction of the muscle.
2. Attitude of the patient—By seeing the attitude or decubitus, one can say that flexor tone has increased in the upper extremity and extensor tone has increased in the lower extremity on the affected side in a hemiplegic patient.
3. Hypotonic muscles are abnormally soft and flabby on palpation.
4. When lifted up and released, a hypotonic limb falls like a log of wood (i.e., it behaves in such a way as if the limb does not belong to the patient).
- 5 Ask the patient to outstretch the upper limbs and spread the fingers : Hypotonic limb may assume an abnormal posture i.e., hyperextended at elbow, hyperpronated at forearm, flexed at wrist and hyperextended fingers at MCP joints is known as 'dinner-fork deformity'.

Always compare with the opposite side while assessing tone.

Patient must be fully relaxed and comfortable before assessing tone.

Causes of cord compression at multiple levels :

- | | |
|---|---------------------------------|
| 1. Neurofibromatosis. | 4. Secondary deposits. |
| 2. Patchy arachnoiditis. | 5. Cervical spondylosis. |
| 3. Prolapsed intervertebral disc (PID). | 6. Arteriovenous malformations. |

Causes of pure motor paraplegia :

- | | |
|-----------------------------------|---|
| 1. Hereditary spastic paraplegia. | 4. Amyotrophic lateral sclerosis (MND). |
| 2. Lathyrism. | 5. Fluorosis. |
| 3. G. B. syndrome. | 6. Erb's spastic paraplegia (syphilitic). |

Recognition of minor weakness in a limb :

By early and easy fatiguability.

Sudden onset of 'generalised weakness' without

1. Transient ischaemic attack (TIA).
2. Hypoglycaemia (subjective weakness).
3. G. B. syndrome.
4. Acute transverse myelitis.
5. Acute anterior poliomyelitis.
6. Poisoning (botulinum).

loss of consciousness :

7. Electrolyte imbalance (1K⁺, iCa²⁺, TMg²⁺).
8. Snake bite (Elapidae group).
9. Myasthenic crisis.
10. Periodic paralysis.
11. Sleep paralysis/cataplexy.
12. Hysterical.

Patient with hypoglycaemia may go into coma, if not diagnosed in time.

Possible causes of 'funny turns' :

Episodes of lost or altered consciousness may be called as 'black-out', 'funny turns' or 'going dizzy'.

- | | |
|----------------------------|----------------------|
| 1. Hypoglycaemia. | 4. Narcolepsy. |
| 2. Temporal lobe epilepsy. | 5. Hyperventilation. |
| 3. Phobic anxiety states. | 6. Amnesic episodes. |

Spastic paraplegia with loss of deep reflexes :

1. In shock stage.
2. Sometimes seen in the presence of bed sore, urinary tract infection (UTI) or malnutrition—the reflex arc may be suppressed leading to loss of jerks.
3. Haematomyelia (sudden bleeding from a vascular malformation)/myelomalacia (thrombosis of a spinal artery with infarction).
4. Associated radiculitis.
5. INH therapy in caries spine causing peripheral neuropathy.
6. If contracture is developed in lower limbs.

* Bed sore, UTI or malnutrition may result in persistent flaccid paraplegia in UMN lesions.

Causes of quadriplegia :

Weakness of all the four limbs can occur in the lesion from cortex to C₅ level of spinal cord :

- | | |
|---|---------------------------------|
| 1. Cerebral palsy. | 7. Motor neurone disease. |
| 2. Bilateral brainstem lesion (glioma). | 8. G.B. syndrome. |
| 3. Cranio-vertebral anomaly. | 9. Peripheral neuropathy. |
| 4. High cervical cord compression. | 10. Myopathy or polymyositis. |
| 5. Multiple sclerosis. | 11. Myasthenia gravis (crisis). |
| 6. Acute anterior poliomyelitis. | 12. Periodic paralysis. |

* For monoplegia, read the section on 'Hemiplegia'.

What are cranio-vertebral anomalies ?

These are the anomalies around the cranio-vertebral junction, some of which may not produce any disability in life. They are :

1. Platybasia (increase in the basal angle of the skull).

2. Basilar invagination (elevation of floor of posterior fossa of the skull).
3. Atlanto-axial dislocation.
4. Occipitalisation of atlas.
5. Klippel-Feil anomaly (fusion of cervical vertebrae).
6. Arnold-Chiari malformation (medulla and cerebellum are extended downwards through foramen magnum).
7. Hypoplastic or separate odontoid process.

The usual **manifestations** of cranio-vertebral anomaly are :

1. Short neck, restricted cervical movements and low posterior hairline (Feil's triad).
2. Spastic quadriparesis of gradual onset; hypertonia, brisk jerks in all the limbs, presence of Babinski's sign.
3. Wasting of the small muscles of hands.
4. Cerebellar signs.
5. Mirror-image movement, impaired position and vibration sense etc.

* Low hairline = hairline below C₄ vertebra.

** Short neck = Ratio of length of the body to length of the neck is > 13 : 1.

Features of 'high' cervical cord lesion :

High cervical cord lesion is seen in cranio-vertebral anomaly, fracture-dislocation of upper cervical spine, haematomyelia, cervical spondylosis and cervical cord tumours. The common features are :

1. Horner's syndrome.
2. Features of injury to spinal accessory nerve (paralysis of sternomastoid and trapezius).
3. Features of injury to spinal tract of trigeminal nerve (i.e., analgesia and thermo-anaesthesia may be limited to ophthalmic and maxillary division in face).
4. Vertical nystagmus (down-beating).
5. Mirror-image movement (sometimes seen in Arnold-Chiari malformation).

* Plus there is quadriplegia.

Possible aetiology of episodic paralysis :

1. Myasthenia gravis.
2. Hyperthyroidism. ■
3. Periodic paralysis (hypokalaemic, hyperkalaemic, normokalaemic).
4. Botulinum poisoning.
5. Primary hyperaldosteronism.
6. G. B. syndrome.

What is lathyrism ?

It is a slowly evolving epidemic pure motor paraplegia (spastic) due to consumption of 'Khesari dal' (lathyrus sativus; a drought-resistant pulse) for long period. Ultimately, the patients develop into disabling spastic paraplegia when they have to crawl on hands. It is common in U.P, Bihar, Rajasthan and M.P in India, and many persons either of the same family or of the same locality may be affected. The causative factor is a neurotoxin and is known as BOAA (Beta Oxalyl Amino Alanine).

What is arachnoiditis ?

A non-specific term referring to inflammation of arachnoid, usually of unknown origin, which may result from tuberculosis, syphilis, sarcoidosis or a post-operative event (e.g., spinal surgery). Constant and severe bilateral asymmetric radicular limb pain is characteristic. Features of radiculopathy is pronounced than features of spinal involvement. Slight fever, features of root compression (e.g., areflexia) may be present. Lumbosacral region is commonly affected. Lumbar puncture may reveal partial or total spinal block. No satisfactory treatment is available.

What is tropical spastic paraplegia ?

It is the HTLV-I associated non-compressive myelopathy where the patient (commonly women) at her third to sixth decade of life complains of gradual onset of weakness of legs (paraparesis), and usually confined to wheel-chair within 10 years. Signs of UMN lesion are found in legs; sensory symptoms and signs are rarely obtained. Bladder involvement with constipation are common. Usually the patient needs myelogram or MRI scan to exclude compressive myelopathy. Affected persons become seropositive for HTLV-I. Few patients show minor response to corticosteroid therapy.

Possible causes of paraplegia with optic atrophy :

The second cranial nerve should always be examined in paraplegia. Optic atrophy is found in :

1. Hereditary—Friedreich's ataxia.
2. Demyelinating—Multiple sclerosis, Devic's disease.
3. Infections like tuberculosis, arachnoiditis, syphilis.
4. Subacute combined degeneration, pellagra.
5. Eale's disease.
6. Subacute myelo-optic neuropathy (SMON). alcohol-induced.

Why do you say compressive myelopathy in this patient ?

The features of **cord compression** are :

1. Usually of gradual onset but may be sudden.
2. Presence of root pain.
3. Symptoms start in one leg and then gradually involve the other leg. Both sensory and motor symptoms are present, and there is progressively increasing course of the disease.
4. Asymmetrical signs and symptoms between two lower limbs are very suggestive.
5. Spastic paraplegia with loss of superficial and deep sensation in varying degree and combination. There is presence of girdle-like sensation in the trunk with zone of hyperaesthesia above.
6. Bladder disturbance is common.
7. Deformity or tenderness in the spine goes in favour of cord compression.

All the above mentioned features are present in this patient.

How caries spine causes paraplegia ?

1. Compression of the spinal cord by cold abscess (extradural)—Commonest.
2. Tuberculous myelitis (involvement of the spinal cord from the bone).
3. Tuberculous endarteritis or thrombosis of the radicular artery—Responsible for the sudden onset of paraplegia or acute exacerbation of existing paraplegia.
4. Pachy meningitis or arachnoiditis.
5. Granulation tissue encircling the dura matter.
6. Compression by sequestered bone.
7. Dislocation of vertebra—Rare.
8. Combination of multiple factors.

*** Caries spine should always be thought as the first cause of paraplegia in India, irrespective of age and sex of the patient. Mid-thoracic region is the commonly involved site.**

Can it be a case of metastasis in the spine ?

'Metastasis in the spine' is unlikely in this case as :

1. Age of the patient is only 28 years (though leukaemic and lymphomatous deposits may be seen in the young).
2. Commonly there is sudden onset of paraplegia though gradual onset paraplegia may be seen.
3. No evidence of primary growth is found (like bronchogenic carcinoma, gastric carcinoma, prostatic carcinoma). No lymphadenopathy is detected.
4. If I am allowed to do the X-ray of the spine, it would show that there is (caries spine) diminution of intervertebral space, collapse of the adjacent vertebrae and paravertebral cold abscess ('Metastasis in the spine' shows destruction of the vertebra without any narrowing of the intervertebral disc space).

*** Root pain is more common in malignancy than in caries spine. Tenderness and deformity of the spine are present in both though the tenderness is more common in malignancy. Metastasis causes paraplegia by (i) Compression by the deposits, and (ii) Collapse of the vertebra.**

Synopsis of bladder dysfunction in neurological diseases :

The conscious control of micturition have its centre within pre-frontal cortex. The urinary bladder receives nerve supply from sympathetic (L_1 – L_3 — the nerve of filling) and parasympathetic (S_2 – S_4 — the nerve of evacuation) nerves. Bladder dysfunction arising out of neurological disorders is known as **neurogenic bladder**.

(A) UNINHIBITED (CORTICAL) BLADDER — Found in frontal lobe tumours, parasagittal meningioma, dementia. There is urgency at low bladder volume (like a child) with sudden uncontrolled evacuation of urine in inappropriate time and place. It is also known as 'mental incontinence' (loss of social control of micturition).

(B) SPINAL BLADDER (with or without autonomic dyssynergia)—

- a) Incomplete lesion :
 - (i) Precipitancy—Due to involvement of inhibitory fibres e.g., multiple sclerosis.
 - (ii) Hesitancy—Due to involvement of facilitatory fibres e.g., incomplete cord compression.
- b) Complete lesion :
 - (i) Retention of urine with overflow incontinence — Commonly seen in ‘neural shock stage’ of acute transverse myelitis. Evacuation of the bladder is always incomplete.
 - (ii) AUTOMATIC BLADDER (UMN bladder)—The evacuation is always complete. It is commonly seen when the ‘neural shock stage’ is over and evacuation occurs by local reflex arc. The bladder is small, spastic and the patient complains of frequency, urgency and urge incontinence.
- c) Lesion in the local reflex arc —
 - (i) Sensory paralytic bladder—There is loss of awareness of fullness of bladder e.g., tabes dorsalis, diabetes mellitus, multiple sclerosis. Large volume of urine collects in the bladder with a huge residual volume.
 - (ii) Motor paralytic bladder—Inability to initiate and continue micturition. It is commonly seen in pelvic neoplasm, trauma, polyradiculopathy etc.
 - (iii) AUTONOMOUS BLADDER (LMN bladder)—Commonly seen in cauda equina lesion, pelvic malignancy, spina bifida etc. There is no sensation of bladder fullness but having continual dribbling. UTI is very common.

* Symptoms of neurogenic bladder are urgency, hesitancy, precipitancy, incontinence and retention of urine. Other causes of hesitancy and precipitancy are CVA, head injury, cerebral tumour, BHP, bladder neck obstruction etc.

** **Urinary incontinence** (involuntary urinary leak) is of four types :

Urge incontinence (uncontrolled passage of urine preceded by strong urge to void—due to overactivity of detrusor muscle; may have UTI), stress incontinence (urinary leak after stress i.e., coughing, sneezing), overflow incontinence (urinary leak due to chronic urinary retention), and true incontinence (voids urine at any time and at any position due to loss of sphincteric function).

How do you like to investigate paraplegia ?

1. Blood (R/E)— TC, DC, ESR; sugar, urea and creatinine.
2. Urine—Routine examination and culture-sensitivity test are done.
3. Mantoux test—Positive in tuberculosis and may be negative in lymphoma.
4. Chest X-ray—To exclude tuberculosis, bronchogenic carcinoma, lymphoma or metastasis.
5. X-ray **of the spine** (antero-posterior and lateral view)—Helps in the diagnosis of caries spine, metastasis, fracture or dislocation of vertebra. It helps in detection of the site of spinal lesion.
6. Lymph node biopsy (if possible).
7. CSF examination—Features of the Froin’s loculation syndrome (below the level of cord compression) are :
 - (i) Low CSF pressure.
 - (ii) Xanthochromia.
 - (iii) Clot formation on standing.
 - (iv) High protein content.
 - (v) Positive Queckenstedt’s test (i.e., no rise in CSF pressure following compression of the internal jugular vein).

CSF examination may also be helpful in multiple sclerosis, arachnoiditis and G.B. syndrome. For the CSF findings in G. B. syndrome, read the section on ‘Peripheral neuropathy’.

8. Myelography (by myodil or metrizamide)—Helps in detection of the level and nature of obstruction. Many a time, combined **CT myelography** is done. Now-a-days, it has been replaced by **MRI scan** of the spinal cord.
9. Miscellaneous—Muscle biopsy, muscle enzymes, EMG (demyelination in G. B. syndrome), blood for vitamin B₁₂ estimation, VDRL and Kahn test, blood for HIV (these tests are done for non-compressive myelopathy), or albumino-cytological dissociation in G. B. syndrome.

* Froin’s loculation syndrome usually results from spinal block as a result of spinal tumour.

Common causes of xanthochromia :

'Yellowish discolouration of the CSF' (xanthochromia) is due to :

1. Old subarachnoid haemorrhage (due to presence of old blood).
2. Guillain-Barre syndrome (due to high protein).
3. Acoustic neurofibroma (due to high protein).
4. Froin's loculation syndrome (spinal subarachnoid block and is due to high protein).
5. Deep jaundice (due to high bilirubin).
6. Massive old intracerebral bleed or haemorrhagic infarction (due to presence of old blood).

Causes of albumino-cytological dissociation in CSF :

It means there is increased protein in CSF without any rise in cell count. The causes are :

1. Guillain-Barre syndrome.
2. Froin's loculation syndrome.
3. Acoustic neurofibroma.

Complications of paraplegia :

1. Pressure sores (trophic changes).
2. Urinary tract infections.
3. Faecal impaction.
4. Contracture of limbs.
5. Chronic renal failure (a common cause of death).

How do you like to manage paraplegia :

1. Nutritious diet—In an adult, 3500 calories/day should be given. There is no need to put the patient on Ryle's tube. Arrange for an alarm bell at the bedside.
2. **Care of the bladder, bowel and trophic ulcers—**
 - a) Bladder—Put a self-retaining catheter under aseptic technique, change the catheter regularly at 2-3 weeks interval, bladder wash, to help in bladder control by application of clip to the drainage tube, R/E and C/S of urine—as and when necessary, antibiotics in UTI.
 - b) Bowel—Treatment of constipation is done by laxatives; manual evacuation may be necessary. Rectal incontinence is difficult to treat.
 - c) Skin—Bed sores are formed due to loss of sensation and diminished blood supply. So the patient should be re-positioned every 2-4 hourly (change of posture) and the skin should be kept dry and clean. Specialised beds (air or water cushioned bed), ripple mattresses, and soft bed coverings prevent development of bed sores. Trophic ulcers (if formed) are cleaned by hydrogen peroxide, dressed by sofra tulle. Aseptic care should always be taken. Plastic surgery - if necessary.
3. Muscle spasms are treated by diazepam, baclofen (5 mg, thrice daily, orally), tizanidine (2mg, thrice daily, orally) or botulinum toxin.
4. The underlying cause should be treated e.g.,—
 - a) In caries spine—Application of traction in early stage. Later on 'plaster jacket' is applied for immobilisation. Antituberculosis chemotherapy is usually continued for at least 9 months to 1 year.
 - b) Treatment of carcinoma or lymphoma by radiotherapy and/or chemotherapy.
 - c) Acute transverse myelitis—High dose corticosteroids (1 mg/kg/day) for two weeks with gradual tapering; high dose methyl prednisolone is very effective.
 - d) G. B. syndrome—Read the last question in section on 'Peripheral neuropathy'.
5. Physiotherapy—Aim is to obtain the maximum development of all those muscles in which voluntary power remains and prevention of flexor contractures in the lower limbs. Passive movements are carried out in the lower limbs once or twice daily. Later on arrangement of wheel chair, walking callipers are done according to the necessity (rehabilitation).
6. Surgery—Drainage of cold abscess, fusion of the vertebra, laminectomy for caries spine, skin grafting for bed sore are rarely needed.

* **Acute transverse myelitis and G.B.syndrome are given as long cases.**

- (iv) Power—
Lower limbs—Proximal muscles are of grade V but extensors of legs are of Grade II; flexor of legs are of Grade III.
Upper limbs—Proximal muscles are of Grade V but the distal muscles are of grade IV.
No involvement of trunk or respiratory muscles.
- (v) Coordination —
Upper limbs—Within normal limit.
Lower limbs—Could not be tested.
- (vi) Involuntary movements—Nil.
- g) Sensory functions—
1. Superficial—Pain, temperature and touch sensations are lost in the distal part of the limbs and it is rather bilaterally symmetrical involvement. Proximal sensations are relatively intact. No involvement of face and neck, and the sensory involvement is 'glove and stocking' in distribution (there is no 'definite' line of demarcation between normal and anaesthetic skin); patchy involvement.
 2. Deep—
 - (i) Vibration sense—Lost distally, impaired proximally.
 - (ii) Muscle sense—Calf muscles are tender on palpation and squeezing (Abadie's sign).
 - (iii) Position sense—Lost distally.
 - (iv) Joint sense—Lost distally.
 3. Cortical—
 - (i) One point localisation ^ 1
 - (ii) Two point discrimination
 - (iii) Stereognosis 1
 - (iv) Graphaesthesia
 - (v) Sensory extinction *

f All are impaired

1

* In a patient of peripheral neuropathy, cortical sensations can not be tested properly,

h) Reflexes—

- | | Right | Left |
|--|-----------------------|--------------------|
| 1. Superficial— | | |
| (i) Abdominal | No response | No response |
| (ii) Cremasteric | No response | No response |
| (iii) Plantar response | No response | No response |
| 2. Deep— | | |
| (i) Biceps jerk | Normal | Normal |
| (ii) Triceps jerk | Normal | Normal |
| (iii) Supinator jerk | Diminished | Diminished |
| (iv) Knee jerk | Diminished | Diminished |
| (v) Ankle jerk | Lost | Lost |
| (vi) Clonus | Absent | Absent |
| 3. Visceral— | | |
| (i) Bladder | 1 | |
| (ii) Bowel | f Within normal limit | |
| (iii) Swallowing |) | |
| 4. Other reflexes—Glabellar tap, grasp reflex and palmo-mental reflex revealed no abnormality. | | |

* *Plantar response is never extensor in peripheral neuropathy.*

- i) Trophic changes—A trophic ulcer is seen in the ball of the great toe of right foot; a large bed sore is seen over the sacrum. Charcot joint is not present,
- j) Cerebellar functions—Normal.
- k) Autonomic functions—Within normal limit (important in diabetes mellitus and G.B. syndrome).
- l) Gait—Could not be tested (there will be high-stepping gait if the patient could walk).
- Romberg's sign—Could not be tested.

(C) CVS :

- (i) Pulse and BP—Describe the pulse in details; BP is 140/80 mm of Hg (supine).
- (ii) Neck veins—Neither engorged nor pulsatile.
- (iii) Shape of precordium—Within normal limit.
- (iv) Apex beat—Present in left 5th ICS, 1 1/2" inside the left MCL.
- (v) Pulsation in different areas—No pulsation seen.
- (vi) Palpation of different areas—No thrill, no pulsation felt, no palpable heart sound.

Case 18

PERIPHERAL NEUROPATHY

What is your diagnosis ?

This is a case of peripheral neuropathy resulting from chronic alcoholism.

How your patient presented himself ?

The 55 years male patient presented in the hospital 2 weeks back with the chief complaints of :

1. Sensation of pins and needles (tingling and numbness) in the distal parts of the upper and lower limbs for last 5 months, with
2. Loss of power or weakness in all the four limbs for last 3 months which was more pronounced distally, and
3. Wasting (thinning) of the distal part of the limbs with pain in the limbs for last 1 month.

The onset of the disease was insidious and it was gradually progressive. The tingling sensation started distally and it was limited in the 'glove and stocking' distribution (feet earlier than hands). The weakness started in the distal part of all the four limbs on different dates. According to the patient, there was no weakness in the proximal muscles and the weakness was not related to repetitive use of muscles (vide myasthenia gravis). He also complained of weakness in the grip and difficulty in holding objects. However, according to the patient, legs are weaker than hands. There was no H/O waxing and waning (vide multiple sclerosis). He had no H/O respiratory distress (vide G.B. syndrome). He is now facing difficulty in standing and walking. The patient is not complaining of any bladder and bowel symptoms at present.

He did not suffer from haematemesis or melaena (due to alcoholism), headache, vomiting, convulsions, speech difficulty, diplopia, nasal regurgitation or intonation. There was neither any past H/O spinal trauma, fever, H/O contact with tuberculosis, H/O exposure to STD, vaccination in recent past, exposure to arsenicals or INH or vincristine, nor the patient was suffering from **diabetes mellitus**. The patient was taking alcohol in a dose of 60-90 ml of whisky, more or less, daily for last 15 years.

* Preceding the 'glove and stocking' type of paraesthesia, there may be hyperaesthesia. The distal paraesthesia usually affects the feet first and then the hands are affected.

Special points in the physical examination in peripheral neuropathy :

(A) GENERAL SURVEY :

- a) The patient is conscious and co-operative.
- b) Anaemia—Moderate.
- c) Jaundice—Absent.
- d) Oedema—Nil.
- e) Parotid glands—**A** bit swollen (due to chronic alcoholism).
- f) Gynaecomastia—Absent.
- g) Facies—A bit flushed and anxious (due to chronic alcoholism).
- h) BP—140/80 mm of Hg.
- i) Skin—No white spots detected (vide leprosy).
- j) All the limbs are lying helplessly with bilateral foot drop.
- k) Ulnar and common peroneal nerves—Not thickened (vide leprosy).
- l) Decubitus—Dorsal decubitus at present; lying helplessly with bilateral foot drop

* Mention all the points in general survey.

(B) NERVOUS SYSTEM :

- a) The patient is conscious, alert, oriented and co-operative.
- b) Higher functions—Normal; speech is not affected.
- c) Cranium and spine—Within normal limit.
- d) Neck rigidity, Kernig's sign—Absent; straight leg raising test—negative.
- e) Cranial nerves—There is neither palatal palsy (diphtheria) nor Vllth nerve palsy (G.B. syndrome). Ophthalmoscopy—Not done.
- f) Motor functions—
 - (i) Attitude—Bilateral foot drop with paucity of spontaneous movements in the limbs.
 - (ii) Nutrition—Wasting of small muscles of the feet and hands. Wasting is clearly seen in extensors of legs.
 - (iii) Tone—Diminished in all the limbs, specially in the distal muscles.



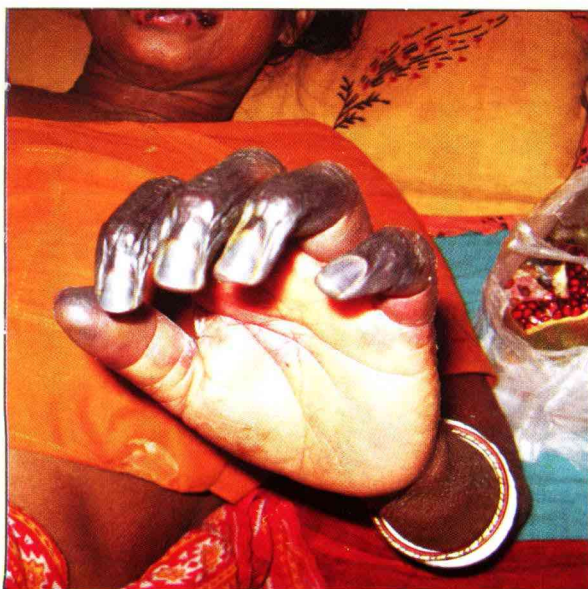
Claw hand deformity



Pes cavus (claw foot). The foot is high-arched; callosity has developed in the sole



Flat foot deformity (pes planus) in Marfan's syndrome



Dry gangrene of the digits. This patient of septicaemia had gangrene of all the digits of four limbs – 'symmetrical peripheral gangrene'



Diabetic foot in a patient of irregularly treated T₂DM



Infiltrated ear in **lepromatous leprosy**



Acute swelling of left first metatarsophalangeal joint in **gout** – hot, red, swollen, painful joint with shiny overlying skin



Gynaecomastia (unilateral) developed after spironolactone therapy in cirrhosis of liver. Venous engorgement in the abdomen due to portal hypertension is seen



Locomotor brachialis seen in a patient of thrombotic stroke



Visible, inflamed and tender temporal artery in **temporal arteritis** (giant cell arteritis); the patient presented with severe headache and claudication of the jaw

(vii) Heart sounds—Within normal limit.

(viii) Murmur—Absent.

D) G. I. TRACT AND GENITOURINARY SYSTEM :

(i) Angular stomatitis—Present.

(ii) Glossitis—Present.

(iii) Epigastrium—Tender on palpation.

(iv) No venous prominence over abdomen.

(v) Spleen—Not palpable.

(vi) Liver—Mild enlargement, soft and tender.

(vii) Ascites—Absent.

(viii) Testes—Within normal limit.

(ix) P/R examination—Not done.

* All the G.I. features are due to chronic alcoholism (vitamin and iron deficiency).

(E) RESPIRATORY SYSTEM :

(1) Shape of the chest

^

(ii) Movement of the chest

I Within normal limit

(iii) Trachea and apex beat

J

(iv) Percussion—Normal resonant note on both sides of the chest.

(v) Breath sound—Vesicular.

(vi) Vocal resonance—Normal on both sides.

(vii) Adventitious sound—None.

(F) LYMPHORETICULAR SYSTEM :

(i) Lymphadenopathy—Absent.

(ii) Sternal tenderness—Absent.

(iii) Haemorrhagic spots—Absent.

Why do you say peripheral neuropathy ?

Discuss the presenting symptoms plus important signs elicited. Give stress on power of muscles, details of sensory functions and trophic changes.

Importance of 'past history' in peripheral neuropathy :

1. H/O chronic alcohol intake.
2. Diabetes mellitus, renal failure or leprosy.
3. Spinal trauma.
 - 4 H/O contact with tuberculosis, vaccination in recent past, H/O exposure (HIV infection).
5. Exposure to drugs (INH, vincristine), solvents, pesticides or heavy metals.
6. Fever (areflexic paralysis in G.B. syndrome).

Common causes of peripheral neuropathy :

(A) Toxic—

1. Alcohol.
2. Pb, As, Hg, TOCP poisoning.
3. Drugs—Vincristine, nitrofurantoin (NFT), INH (by causing pyridoxine deficiency).
4. Radiation.

(B) Metabolic—

1. Diabetes mellitus.
2. Chronic renal failure (CRF).
3. Porphyria.
4. Hypothyroidism.
5. Amyloidosis.

(C) Injective or inflammatory—

1. Leprosy.
2. Diphtheria.
3. Landry-Guillain-Barre syndrome.
4. Viral hepatitis.
5. Enteric fever.
6. HIV infection.

(D) Deficiency—Thiamine, nicotinic acid, pyridoxine, vitamin B₁₂, pantothenic acid, vitamin E and iodine

(E) Connective tissue diseases—

1. SLE.
2. Rheumatoid arthritis.
3. Polyarteritis nodosa.
4. Cryoglobulinaemia.
5. Sjogren's syndrome.

(F) Disseminated malignancy, paraneoplastic syndrome, lymphoma, multiple myeloma.**(G) Hereditary or idiopathic (e.g., Charcot-Marie-Tooth disease).**

Pathogenesis of peripheral neuropathy is classified into two types : demyelination and axonal. Acquired demyelinating polyneuropathy falls into two major groups : acute form, i.e., G. B. syndrome (AIDP), and more chronic forms, i.e., chronic inflammatory demyelinating polyneuropathy (CIDP, steroid-dependent).

**** Small fibre neuropathy** : muscle power and deep reflexes are less affected than the affection of pain and temperature sensations.

Large fibre neuropathy : significant motor disturbance and loss of deep reflexes with minimal sensory loss.

Five common causes of peripheral neuropathy in clinical practice :

1. Leprosy (commonest cause globally).
2. Diabetes mellitus.
3. G.B. syndrome.
4. Chronic renal failure (CRF), and
5. Toxin-induced : alcohol-induced or INH therapy.

Classification of neuropathy by evolution :

- Acute : G. B. syndrome, diphtheria, porphyria, malignancy, drugs (TOCP, As)
- Subacute : alcoholic, nutritional, toxic (hexacarbon)
- Chronic : diabetes, CRF, CIDP, paraneoplastic
- Longstanding : Charcot-Marie-Tooth disease
- Recurrent : relapsing CIDP, porphyria, occupational toxic neuropathy, alcoholic

Sensations carried by long tracts :**a) The spinothalamic tract (lateral column) carries :**

- (i) Pain.
- (ii) Temperature (hot and cold), and
- (iii) Crude touch.

Lateral column also contains pyramidal tract (motor pathway).

The tract of Goll and Burdach (posterior column) carries :

- (i) Vibration sense
- (ii) Muscle sense
- (iii) Pressure sense, Proprioception or deep sensation
- (iv) Joint sense
- (v) Position sense
- (vi) Fine touch (well-localized touch), and
- (vii) Cortical sensations (assay parietal lobe functions)

* Testing of sensory functions are described in the section on 'Charcot joint'.

Peripheral neuropathy with hypertension :

- | | |
|--------------------------|--|
| 1. G. B. syndrome. | 4. Chronic renal failure. |
| 2. Polyarteritis nodosa. | 5. Porphyria (acute Intermittent variety). |
| 3. Lead poisoning. | g. Amyloidosis. |

Predominantly motor neuropathy :

- | | |
|--------------------------------|-----------------------------|
| 1. G. B. syndrome. | g Infectious mononucleosis. |
| 2. Porphyria. | 7. Lead poisoning. |
| 3. Connective tissue diseases. | 8. HIV/AIDS |
| 4. Diphtheria. | g Hereditary neuropathy. |
| ^ . . Paraneoplastic. | 10. Dapsone-induced. |

^{r,athThefe} <causes of Proximal motor weakness > distal motor weakness in peripheral neuropathy, also in diabetic amyotrophy, proximal muscles are predominantly affected.

Predominantly sensory neuropathy :

1. Leprosy.
2. Drugs (vincristine, NFT, INH).
3. Diabetes mellitus.
4. Renal failure (initially).
5. Hereditary sensory neuropathy.
6. HIV infection.
7. Paraneoplastic syndrome.
8. Thalidomide toxicity.

Peripheral neuropathy with cranial nerve involvement :

1. G. B. syndrome (VII): Miller Fisher syndrome (III, IV).
2. Leprosy (VII).
3. Diabetes mellitus (III, VI).
4. Diphtheria (III, VI, IX, X).
5. Sarcoidosis (VII).

Painful peripheral neuropathy :

1. Diabetes mellitus.
2. Alcoholic polyneuropathy.
3. Carcinomatous neuropathy.
4. Vitamin B, and B₁₂ deficiency.
5. Porphyria.
6. Arsenic-induced.

Drugs causing peripheral neuropathy :

1. INH
2. Nitrofurantoin (NFT).
3. Vincristine.
4. Paclitaxel.
5. Thalidomide.
6. Cisplatin.
7. Suramin.
8. Amiodarone.
9. Pentamidine.
10. Pyridoxine (high dose).
11. Stavudine.

Causes of mononeuritis multiplex :

It is the asymmetrical involvement of multiple non-contiguous peripheral nerves serially or at a time over days to years, and is usually due to arteritic involvement of vasa nervorum (i.e., multiple mononeuropathy). Diagnosis is done clinically and is supported by electrical studies (nerve conduction velocity). It gives a picture of distal symmetric neuropathy and is commonly found in :

1. Leprosy.
2. Diabetes mellitus.
3. Polyarteritis nodosa.
4. Sarcoidosis.
5. Rheumatoid arthritis.
6. Amyloidosis.
7. Vasculitis.
8. Trauma.
9. HIV infection.
10. G. B. syndrome.

The involvement is usually asymmetrical with predominant affection of the lower limbs.

N.B. : • Neuropathy—pathological process affecting peripheral nerve or nerves.

- Peripheral neuropathy or polyneuropathy—a diffuse symmetrical disease process of peripheral nerves (peripheral means that the disorder is outside the CNS) involving the peripheral parts of body and gradually progresses proximally.
- Mononeuropathy—focal involvement of a single nerve trunk (e.g., ulnar, radial or median nerve).
- Mononeuritis multiplex—already described above.
- Radiculopathy—a pathological process which affects the nerve roots.

Which distal sensation is lost early in peripheral neuropathy ?

Vibration sense is lost early in peripheral neuropathy (very common feature in diabetic neuropathy). Peripheral neuropathy follows the rule—'the longer the nerve fibre (i.e., axon length), the earlier is the involvement', and this is why the distal part of the body ('glove and stocking' distribution) is affected earliest and most.

How diphtheritic neuropathy presents ?

1. Paralysis of the soft palate and posterior pharyngeal wall may occur very early (within 10 days).
2. Then paralysis of accommodation follows (difficulty in reading small objects).
3. Polyneuropathy usually develops 2-6 weeks after the onset of the disease. Weakness involves all the four limbs and descends from arms to legs.

* Among all cranial nerves the IIIrd, Vth, IXth and Xth nerves are commonly affected.

Presentations of diabetic neuropathy :

1. Distal, symmetrical, mixed sensory-motor polyneuropathy (commonest)—mainly small fibre, mixed type or large fibre neuropathy.
2. Asymmetrical, proximal motor neuropathy (diabetic amyotrophy).
3. Distal, symmetrical, sensory polyneuropathy.
4. Asymmetric mononeuritis multiplex (painful).
5. Acute mononeuropathy (isolated, painless IIIrd, IVth and VIth cranial nerve palsy; affection of ulnar, median, femoral nerves rarely).
6. Radiculopathy (usually involves spinal nerves, over the chest wall or abdomen).
7. Autonomic neuropathy.

Neuropathy with predominant upper limb involvement :

1. Diphtheria.
2. Porphyria.

Peripheral neuropathy with significant autonomic involvement :

- | | |
|----------------------------------|------------------------|
| 1. Diabetes mellitus. | 5. AIDS. |
| 2. G. B. syndrome. | 6. Amyloidosis. |
| 3. Acute intermittent porphyria. | 7. Botulism. |
| 4. Peroneal muscular atrophy. | 8. Sjogren's syndrome. |

Table 14 : Differentiation between myelopathy, neuropathy and myopathy

Features	Myelopathy	Neuropathy	Myopathy
1. Onset	1. Any age	1. Any age	1. Childhood
2. Tingling and numbness	2. Very rare	2. Present	2. Never occurs
3. Bladder & bowel involvement	3. Occurs	3. Usually absent	3. Never occurs
4. Atrophy and pseudohypertrophy	4. Only atrophy	4. Only atrophy	4. Both present
5. Bilaterally symmetrical involvement	5. May be present	5. Initially asymmetrical	5. Present
6. Cranial nerve involvement	6. None	6. May be present (G.B. syndrome and diphtheria)	6. Never occurs
7. Girdle-like sensation	7. Present in compressive myelopathy & acute tr. myelitis	7. Absent	7. Never occurs
8. Sensory involvement	8. Upper border of sensory loss may present	8. Patchy	8. Never occurs
9. Motor involvement	9. According to involvement of myotome	9. Mainly distal	9. Predominantly proximal
10. Tone of muscles	10. Spasticity (except in shock stage)	10. Hypotonia (flaccidity)	10. Hypotonia (terminal stage)
11. Deep reflexes	11. Brisk (except in shock stage)	11. Lost or diminished	11. Usually normal
12. Plantar response	12. Extensor	12. Flexor or no response	12. Flexor
13. Confirmation	13. Myelography, CT. MRI	13. Nerve conduction velocity (NCV)	13. Electromyography (EMG)

Features of root lesion (radiculopathy) :

- (A) Anterior root — Muscle atrophy and weakness in root distribution; rare.
- (B) Posterior root — Root pain, hyperalgesia and hyperaesthesia corresponding to the segments compressed. Different types of sensory abnormalities (e.g., hyperaesthesia) with an upper level which is usually a bit above the segmental level of the site of compression; commonly seen.

What are the differential diagnosis of your case ?**I. Aetiological differentiation :**

- a) *Diabetes mellitus* :
 - 1. H/O polyphagia, polydipsia and polyuria.
 - 2. H/O taking oral hypoglycaemic agents or insulin.
 - 3. Retinopathy invariably present; there may be nephropathy.
 - 4. Susceptibility to infections and delayed wound healing.
- b) *Leprosy* :
 - 1. Typical hypopigmented and anaesthetic skin lesion.
 - 2. Palpable peripheral nerves.
 - 3. Trophic changes.
 - 4. 'Leonine' facies may be present.
- c) *G. B. Syndrome* :
Read the section on 'Paraplegia'.
- d) *Drug-induced* :
 - 1. H/O prolonged intake of INH, NFT or vincristine.
 - 2. Predominantly sensory neuropathy.
 - 3. Presence of primary disease for which the drugs are taken, e.g., tuberculosis, leukaemia etc.
- e) *Arsenical polyneuropathy* :
 - 1. Early sensory symptoms; later on sensory-motor polyneuropathy.
 - 2. Rain-drop skin lesions with hyperkeratosis of palm and sole.
 - 3. People of known geographical area (different districts of West Bengal and Bangladesh) using deep tube-well water are predominantly affected.
 - 4. Mee's line in nail (white transverse banding of nails).
 - 5. Presence of anaemia with or without jaundice (due to hepatic involvement). There may be portal hypertension.
 - 6. Estimation of arsenic in hairs, nails, urine confirms the diagnosis.

II. Similarities in clinical features :

- a) *Tabes dorsalis* :
 - 1. H/O exposure at least 10 years back.
 - 2. Lightning pain; sensation of walking on cotton wool.
 - 3. Muscle sense is lost (calf muscles and even testicles are insensitive on squeezing).
 - 4. Loss of all modalities of posterior column sensations.
 - 5. Tabetic facies; Argyll Robertson pupil and optic atrophy may be seen.
 - 6. Romberg's sign is positive.
- b) *Subacute combined degeneration (SCD)* :
 - 1. Anaemia.
 - 2. Glossitis.
 - 3. Lhermitte's sign—Positive.
 - 4. Ankle jerk — Lost.
 - 5. Plantar response — Extensor.

* SCD is the demyelination of pyramidal tract and posterior column of the spinal cord as a result of vitamin B₁₂ deficiency. Distal paraesthesia, spastic weakness and difficulty in walking are common.

- c) *Myopathy* :
 - 1. History from childhood.
 - 2. No sensory loss.
 - 3. Predominantly proximal muscle weakness.
 - 4. Pseudohypertrophy plus atrophy.
 - 5. No bladder and bowel involvement.
 - 6. Plantar response — Flexor.

What is TOCP neuropathy ?

Triorthocresylphosphate (TOCP) poisoning is also known as 'ginger paralysis'—owing to the consumption of fluid extract of ginger which was used in the manufacture of bootleg alcohol and was adulterated with TOCP. Symptoms of TOCP polyneuropathy usually develops 10-20 days after the consumption of adulterated food (cooking-oil) or drink, and produces bilateral wrist and foot drop with wasting of distal muscles of the limbs.

Causes of foot drop :

It is due to paralysis of the extensors of foot and peronei muscles. The common causes are,

1. Peripheral neuropathy (e.g., leprosy, diabetes mellitus).
2. Common peroneal nerve palsy (commonest cause of unilateral foot drop).
3. Prolapsed intervertebral disc (PID)—Due to lesion in L₅ root.
4. Motor neurone disease.
5. Sciatic nerve lesion.
6. Peroneal muscular atrophy.

* For 'wrist drop', read the section on 'Claw hand'.

Altered muscle sense :

Method : Muscles of calf or forearm are squeezed. Normally patient feels some discomfort.

(A) Diminished :

1. Tabes dorsalis.
2. Syringomyelia.
3. Carcinomatous neuropathy.

(B) Increased :

1. Polyneuropathy (Abadie's sign).
2. Myositis.
3. Subacute combined degeneration.

Investigations you like to perform in peripheral neuropathy ?

1. **Nerve conduction velocity** (lowered in neuropathy but not in myelopathy or myopathy).
2. Nerve biopsy (usually sural nerve is selected at the ankle; specially in inflammatory pathology).
3. Blood for R/E, sugar, urea, creatinine, VDRL, autoantibodies, serum folate and B₁₂, and thyroid and liver function tests.
4. EMG (to differentiate neuropathy from myopathy and diseases of myoneural junction).
5. CSF (done in G.B. syndrome).

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* Electrodagnosis in neurology is done by NCV and EMG. NCV shows features of demyelination or axonal injury; EMG reveals features of denervation.

CSF features in G.B. syndrome :

1. Pressure—Normal.
2. May be xanthochromic.
3. Clots formation on standing.
4. High protein, normal sugar, and
5. No rise in cell count (i.e., albumino-cytological dissociation).

How do you like to manage your patient ?

1. Complete abstinence from intake of alcohol.
2. Rest in bed.
3. Inj. vitamin B₁₂, B₆ and B₁₂ — twice weekly, deep I.M.; inj. methylcobalamin may be tried. For paraesthesia—carbamazepine (300-1200 mg/day), aspirin (350-1200 mg/day) or amitriptyline (25-50 mg/day) may be given. For painful peripheral neuropathy pregabalin, tramadol, gabapentin may be helpful.
4. Physiotherapy and occupational rehabilitation in profound weakness.
5. Splinting of the foot (to manage foot drop); care of the foot-ulcer and bed sore.
6. Empirical treatment with steroid is usually disappointing.

Management of G.B. syndrome :

Treatment should be initiated as early as possible to get the maximum benefit.

1. Intravenous high-dose immunoglobulin therapy (0.4 g/kg/day for 5 days)—total dose 2 g/kg.
2. Plasmapheresis.
3. Physiotherapy.
4. Respiratory assistance in 'Respiratory Care Unit (RCU)' by intubation and positive pressure ventilation when the vital capacity falls below one litre. Good supportive care is given.
5. Corticosteroids or ACTH has shown no beneficial effects; it is better not to use them.

Case 19

PARKINSONISM

What is your diagnosis ?

This is a patient of idiopathic parkinsonism.

* James Parkinson, a physician from London, described the disease in 1817. Idiopathic parkinsonism is also known as 'shaking palsy'.

How your patient presented himself ?

This male patient aged 75 years presented himself with difficulty or paucity of movements (bradykinesia), tremor, and stiffness of limbs and joints (three cardinal symptoms of parkinsonism) for last 3 years. The difficulties started gradually and progressing slowly without any waxing and waning.

1. He is complaining of difficulty in initiation of movement*. To start with there is clumsiness of movement and now he is unable to perform day to day activities. He feels difficulty in getting up from sitting position, walking, doing finer activities like combing or fastening buttons and writing (the letters tend to be smaller at the end of a line — micrographia).
2. Tremor is persisting for last 3 years which is mainly present in hands and occurs mostly at rest (static tremor). Recently the patient has noticed tremor of the legs, jaw and tongue. He has also complained of increasing tremor with emotional stress and diminution temporarily by willed movement (and diminution also during sleep as told him by family members).
3. Rigidity made him stiff and is responsible for his flexed attitude or 'stooped posture'. He knows from his relatives that his voice has turned into slow (dull) and monotonous type. Recently, he is feeling weakness of upper and lower extremities.

He was absolutely normal before 3 years. There is neither any H/O headache, vomiting, convulsions, diplopia, nasal intonation, nasal regurgitation nor there is any bladder and bowel disturbances. He has no complain regarding the sensory system. About the higher function, he is a bit depressed though there is no intellectual deterioration.

* *There is delay in initiation, slowness in expression and paucity in execution in parkinsonism.*

** Parkinsonism is known as akinesia-tremor-rigidity syndrome.

*** *Postural instability*, the most disabling feature, is usually present in advanced disease contributing to falls and resulting injuries.

**** Bradykinesia is common in parkinsonism, myxoedema and depression.

Importance of past, family, personal and treatment history :

(A) PAST HISTORY — H/O

- a) Fever, convulsions, coma (indicates post-encephalitic variety; though there may be a long interval, usually parkinsonism develops during the subsequent 12 months).
- b) Head injury (punch drunk syndrome).
- c) Exposure to STD (neurosyphilis).
- d) Jaundice (Wilson's disease).
- e) High BP (multiple infarction in brain).
- f) CO or Mn intoxication.
- g> Headache, vomiting and convulsions — Brain tumour.

(B) FAMILY HISTORY — H/O hypertension, diabetes mellitus or similar type of illness within the family.

(C) PERSONAL HISTORY — Whether 'boxer' or not.

(D) TREATMENT HISTORY — Whether there is any H/O prolonged intake of phenothiazines, butyrophenones, reserpine etc.

Special points in the physical examination in parkinsonism :

(A) GENERAL SURVEY :

- a) The patient is conscious and co-operative.
- b) Masked facies characterised by,
 - (i) Infrequent blinking with staring look (spontaneous ocular movements are lacking),
 - (ii) Loss of facial expression (looks blank), and
 - (iii) Widened palpebral fissure.

* There may be associated titubation of head.

- c) Pulse — Write all the points. Condition of the arterial wall is normal (excludes atherosclerosis).

- d) BP— 135/85 mm of Hg (excludes atherosclerotic variety).
- e) Jaundice — Absent (for Wilson's disease).
- f) Cornea — K-F ring is absent (for Wilson's disease).
- g) Seborrhoea (greasy skin), sialorrhoea — Absent (excludes post-encephalitic variety).
- h) Outstanding features — Coarse and static tremor of hands; jaw tremor. Rate of eye blinking is reduced to <10/minute.
- i) Posture—flexed posture with impaired postural reflex.

* Mention all the points in general survey.

(B) *NERVOUS SYSTEM* :

- a) Higher functions —
 - (i) The patient is conscious, alert, oriented and co-operative.
 - (ii) Normal intelligence and memory.
 - (iii) A bit depressed.
- b) Speech — Slow, Indistinct and monotonous speech without any fluctuation or modulations (bradylalia as well as hypophonia, and In advanced disease, speech is reduced to muttering).
- c) Neck rigidity and Kernig's sign — Absent.
- d) Cranium and spine — Within normal limit.
- e) Cranial nerves — Normal; Jaw jerk — Normal; oculogyric crisis — Absent.
- f) Motor functions —
 - (i) Attitude — Flexed attitude in all the limbs with a stooped posture.
 - (ii) Nutrition — No wasting seen.
 - (iii) Tone — Increased in all the four limbs; flexor and extensor tone are equally Increased; **rigidity** is mainly of lead pipe (plastic) in type. In the hands (i.e., at wrist joint), they are of cogwheel in type (due to static tremor).
 - (iv) Power—Grade IV in both upper and lower limbs.
 - (v) Coordination — Could not be tested properly.
 - (vi) Involuntary movements — Tremor is present. It is,
 1. **Static tremor** and does not increase at goal-point of an action.
 2. Coarse In type (4-6/second).
 3. More prominent in hands; bilateral and symmetrical.
 4. The patient can control the tremor temporarily by voluntary movement.
 5. There is predominantly contraction of flexor and extensors alternately with a prominent rotatory component between finger and thumb (pill-rolling, bread-cutting or drum-beating tremor); often changing to supination and pronation of forearm.
 6. Tremor is increased by emotion and disappear during sleep.
 7. Tremor is mainly present in the distal part of upper limb; and is also present in the jaw and tongue.
- g) Sensory functions —
 - (i) Superficial
 - (ii) Deep
 - (iii) Cortical

} NAD
- * Mention the subheadings.
- h) Reflexes —
 - (i) Superficial —
 1. Abdominal
 2. Cremasteric
 3. Plantar response

} iWithin normal limit
 - (ii) Deep —
 1. Biceps jerk
 2. Triceps jerk
 3. Supinator jerk
 4. Knee jerk
 5. Ankle jerk
 6. Clonus—Absent

} Within normal limit

- (iii) Visceral —
 - 1. Bladder
 - 2. Bowel, and
 - 3. Swallowing
- } Within normal limit
- (IV) utner renexes —
- 1. **Glabella tap — Present.**
 - 2. Grasp reflex—Absent.
 - 3. Palmo-mental reflex—Absent,

i) Trophic changes — Nil.

j) Cerebellar functions — Normal,

k) Autonomic functions — Within normal limit.

I) Gait — There is ‘festinant’ gait. The features are :

- (i) Stance—Patient bends forward (universal flexion)— known as ‘simian attitude’.
- (ii) Difficulty in initiation of movetnent; the patient may freeze suddenly i.e., glued to the floor.
- (iii) Starts walking with rapid, short, shuffling steps as if the patient is trying to catch up his centre of gravity.
- (iv) Paucity of automatic movements of both the upper limbs (i.e, swinging movement of arms).
- (v) Steps become more and more rapid with a tendency to run (festination) and the patient may ultimately fall like a telegraphic pole.
- (vi) There is presence of propulsion (tendency to fall forward), retropulsion (walking backwards uninhibited, if pulled backwards) and lateropulsion (walking sideways better).
- (vii) Impaired balance on turning.
- (viii) Difficult to stop suddenly if pushed or pulled.
- (ix) Can run faster than walking during emergency (kinesia paradoxa).

(C) CVS — Within normal limit (search for the signs of atherosclerosis).

(D) RESPIRATORY SYSTEM—Within normal limit.

(E) G.I. TRACT AND GENITOURINARY SYSTEM — Normal (search for hepatosplenomegaly due to Wilson's disease, specially in a young patient).

(F) LYMPHOKTICULAR SYSTEM — Normal.

* Mention the subheadings in all the systems.

** Write Plantar response as ‘bilaterally flexor’.

Chief complaints of your patient :

- 1. Difficulty of movements for 3 years,
- 2. Tremor in hands for 3 years,
- 3. Stiffness (rigidity) of muscles for 3 years, and
- 4. Postural instability lor last 2 years.

Causes of masked fades :

See the section on ‘Scleroderma’.

What is glabella tap reflex (Myerson's sign) ?

In health, if a series of sharp finger tap (by index finger) is applied over the glabella (root of the nose), it evokes three or four blinks (bilateral closure of eyes) before the response is inhibited.

Precautions :

- 1. Tap should be light and done at a rate of 1 /second upto 5 taps at a time.
- 2. Tap should be applied on standing behind the patient and the tapping finger is kept above the visual field (so that menace reflex does not play).
- 3. Eyes should be kept open.

The afferent fibres are carried through Vth cranial nerve while the efferent fibres go through VIth cranial nerve, and the centre for this reflex lies in pons.

Prolongation of glabella tap (i.e., eyes continue blinking with each tap) response is seen in parkinsonism, diffuse cortical damage and senile dementia. This reflex tests the integrity of ‘nigro-strial pathway’.

Differentiation between spasticity and rigidity :

Hypertonia is of two types : spasticity and rigidity.

(A) SPASTICITY :

- 1. Alaways in UMN lesion. Spasticity takes some time to develop.

2. Tone is of clasp-knife in type [hypertonia is felt maximally at the beginning of passive movement when the limb is fairly flexed or extended, and the resistance is decreased (gives away) if the movement is continued]—quick movement is better to elicit spasticity.
3. Hypertonia is marked in flexors of upper limbs and extensors of lower limbs, i.e., more in anti-gravity muscles.
4. Jerks or deep reflexes — Brisk; clonus may be present. Abdominal reflexes are lost.
5. Plantar response—Extensor.
6. Involuntary movements are not seen.

* Clasp-knife spasticity is the lengthening and shortening reactions in muscles originally described by Sherrington.

(B) RIGIDITY:

1. Seen in extrapyramidal lesions.
2. Tone is of lead pipe or cogwheel in type—slow movement is better to elicit rigidity.
 - a) Lead pipe — Uniform resistance present throughout the entire range of passive movement, and is mostly appreciated in legs and trunk; also known as plastic type rigidity.
 - b) Cogwheel — Fluctuant resistance to passive movement due to presence of static tremor (as if a lever is rubbing on the teeth of a cogwheel); best observed in wrist joint and neck.
3. Hypertonia is marked in both the limbs equally, i.e., flexors and extensors of all the four limbs are equally affected.
4. Jerks of deep reflexes — Normal; clonus — Absent. Abdominal reflexes are preserved.
5. Plantar response — Flexor.
6. Very often associated with involuntary movements (e.g., tremor).

Aetiology of parkinsonism :

1. Idiopathic (Parkinson's disease or paralysis agitans)—may be due to environmental toxin.
2. Atherosclerotic (vascular).
3. Post-encephalitic (von Economo's disease)—becoming increasingly rare.
4. Punch drunk syndrome in boxers (traumatic or head injury).
5. Wilson's disease.
6. CO, CS₂, methyl-phenyl-tetrahydropyridine (MPTP; used for recreational purposes) and manganese dust intoxication.
7. Neurosyphilis.
8. Drug-induced (reversible parkinsonism) — Phenothiazines, butyrophenones, tetrabenazine, reserpine, metoclopramide.
9. Cerebral tumour (rare).
10. Huntington's disease.

N.B. : 1, 2 and 3 are the most common causes. According to few neurologists, vascular or atherosclerotic variety does not exist, and thus remains a topic of controversy.

Components of extrapyramidal system :

1. Caudate nucleus.
2. Putamen.
3. Globus pallidus.
4. Red nucleus.
5. Substantia nigra.
6. Subthalamic nucleus of Luys.
7. Dentate nucleus (of cerebellum).
8. Reticular formation (of brainstem).

Eye signs in parkinsonism :

1. Infrequent blinking; staring look.
2. Impaired pursuit movement of eyeball.
3. Reflex blepharospasm (Myerson's sign)
4. Oculogyric crisis (post-encephalitic).
5. Reversed Argyll Robertson pupil.
6. On closing the eyes, there is fluttering of eyelids (blepharoclonus).
7. Hypometric saccades.

Symptoms of extrapyramidal diseases ;

(A) HYPERKINESIA :

- | | |
|---|---|
| <ol style="list-style-type: none"> 1. Tremor (static). 2. Chorea. 3. Athetosis. 4. Hemiballismus. | <ol style="list-style-type: none"> 5. Myoclonus. 6. Dyskinesia. 7. Dystonia. |
|---|---|

(B) HYPOKINESIA OR AKINESIA : Poverty and slowness of movement (e.g., parkinsonism).

(C) ATAXIA (POSTURAL DISORDER).

(D) RIGIDITY.

* Hyperkinesia, hypokinesia and akinesia are collectively known as '**movement disorders**'.

What are the stages of parkinsonism ?

Stage I — Unilateral involvement ('hemiplegic' parkinsonism).

Stage II — Bilateral involvement but no postural abnormality.

Stage III — Bilateral involvement with mild postural abnormality.

Stage IV — Stage 3 plus severe postural abnormality requiring substantial help.

Stage V — Severe, fully developed disease; patient is restricted to bed and wheel chair.

What is 'Parkinsonism-plus' syndrome ?

Classical parkinsonian features (akinesia-rigidity) are seen in combination with autonomic, cerebellar, oculomotor and cortical dysfunction in various neuro-degenerative diseases like,

1. Progressive supranuclear palsy (Steele-Richardson syndrome)—commonest.
2. Multisystem degeneration e.g., Shy-Drager syndrome, striatonigral degeneration, olivopontocerebellar atrophy.
3. Wilson's disease.
4. Huntington's chorea.
5. Normal-pressure hydrocephalus.
6. Alzheimer's disease. Pick's disease.

Table 15 : Differentiation between idiopathic and post encephalitic parkinsonism

Features	Idiopathic	Post-encephalitic
1. Age	1. Elderly or late middle-aged	1. Relatively younger
2. Onset	2. Insidious	2. Sudden
3. Past history	3. Nothing particular	3. Gives history of encephalitis
4. Presentation	4. Mainly tremor	4. Rigidity with impaired higher function
5. Higher function	5. Normal	5. Often mentally dull
6. Cranial nerves	6. Normal	6. Oculogyric crisis (involuntary upward conjugate deviation of eyeballs)
7. Rigidity	7. Usually of cogwheel in type	7. Lead pipe in type (due to absence of tremor)
8. Tremor	8. Most prominent feature i.e., tremor > rigidity	8. Less prominent; rigidity is the main feature i.e., rigidity > tremor
9. Reflexes	9. Normal	9. Jerks are brisk and plantar response is extensor due to involvement of pyramidal tract
10. Autonomic features	10. None	10. Flushing of skin, seborrhoea, sialorrhoea (excessive salivation), behavioural disturbances, hiccoughs and orthostatic hypotension
11. L-dopa therapy	11. Good response	11. Poor response

* **Oculogyric crisis** may be seen in drug-induced dyskinesia, petit mal epilepsy and in post-encephalitic parkinsonism. The head may tilt back and body may take a posture of opisthotonus.

Features of atherosclerotic parkinsonism :

1. Age—Usually above 60 years.
2. Higher function —Loss of memory is very common (due to multiple infarction in brain).
3. Bradykinesia is the prominent feature. Tremor and rigidity are less pronounced.
4. Clinical evidences of atherosclerosis (see the section on 'Hemiplegia').
5. Bilateral UMN signs may be seen e.g.,
 - (i) Jaw jerk—Present.
 - (ii) Plantar response—Bilaterally extensor.

6. Course of the disease is more rapid in comparison to other types.
7. Little or no response to L-dopa therapy.

What are the differential diagnosis of your case ?

- (A) Causes of tremor—Read the section on 'tremor'.
- (B) Causes of hypertonia—Read the section on 'Paraplegia'.
- (C) Causes of 'parkinsonism-plus' syndrome (described above).

What is the pathology in parkinsonism ?

Melanin is lost from the dopaminergic nerve cells in substantia nigra, and there is nerve cell loss with gliosis and formation of Lewy body (intracytoplasmic inclusion body). It is seen that the nigro-striatal pathway utilise dopamine as a neurotransmitter and subsequently dopamine loss produces parkinsonism (nerve cell loss elsewhere in the basal ganglia is also responsible for depletion of dopamine). It is a disease of extrapyramidal system (basal ganglia) due either to an increase in acetylcholine or decrease in dopamine.

Investigations you like to perform in a case of parkinsonism :

Diagnosis of parkinsonism is essentially made on clinical grounds though investigations may be required in exceptional situations.

- (A) Blood—Serum copper, ceruloplasmin, VDRL, Kahn test.
- (B) Urine—Urinary copper estimation (in 24 hours).
- (C) Liver function tests (for detection of Wilson's disease).
- (D) CT or MRI scan of brain is done in,
 - (i) Patients < 50 years.
 - (ii) Unilateral disease.
 - (iii) Signs of UMN, cerebellar or autonomic involvement.
 - (iv) Diagnosis in doubt.

* Patients < 50 years of age are screened for Wilson's disease.

Outline of treatment of parkinsonism :

- (A) Stage I and II (mild)—Medication may be required. Only anticholinergic drugs and selegiline are used; amantadine may be tried.
- (B) Stage II, III and IV (moderate and severe) —
 - a) Levodopa, carbidopa, bromocriptine, pergolide, lisuride, pramipexole, ropinirole, cabergoline, different COMT inhibitors (tolcapone, entacapone).
 - b) Stereotactic surgery.
 - c) Physiotherapy and speech therapy.

* Tremor and rigidity are best controlled by anticholinergic drugs. Akinesia and postural instability are improved by levodopa. Selegiline reduces the free-radical formation generated from the oxidation of catecholamines. COMT stands for catechol-o-methyl-transferase; COMT inhibitors (tolcapone, entacapone) enhance the benefits of levodopa therapy and are useful in patients with response fluctuation to levodopa.

** Other drugs used ; rasagiline, rivastigmine, antioxidants (vitamin C and E).

Name some anticholinergic drugs used in parkinsonism :

- 1) Trihexyphenidyl 2) Benztropine 3) Orphenadrine 4) Biperiden 5) Benhexol.

Indications of surgery in parkinsonism :

Stereotactic surgery is done in globus pallidus or ventrolateral thalamus. Indications of operation are (obviously with no response to drugs) :

1. Relatively young patient.
2. Good general health with sound mental health.
3. Unilateral tremor or rigidity (akinesia is less responsive to surgery).

* Neural transplant techniques like adrenal medullary transplant to striatum or glial cell-line neurotrophic releasing factor into cerebral ventricles or basal ganglia are tried; not of much value.

Side effects of drugs used in parkinsonism :

- I. Levodopa—Nausea, vomiting, dyskinesia, postural hypotension, on-off phenomenon, hallucinations.
- II. Anticholinergic drugs—Dry mouth, constipation, blurring of vision, glaucoma, retention of urine, involuntary movements and confusion (use anticholinergic drugs cautiously in aged).

III. Amantadine—Mental confusion, ankle oedema, livido reticularis, seizures, urinary retention.

IV. Selegiline (deprenyl)—Postural hypotension, involuntary movements, psychosis.

* Now-a-days carbidopa (levodopa with DOPA decarboxylase inhibitor in a ratio of 10 :1. DOPA decarboxylase inhibitor prevents destruction of levodopa in the blood stream) is preferred to levodopa. Propranolol is sometimes used to treat tremor.

What is on-off phenomenon ?

After 3-5 years of treatment with levodopa, a short period of dyskinesia may develop (e.g., athetosis of the neck). Akinesia and weakness may last for few minutes to hours and may alternate with dyskinesia. These symptoms are due to narrow therapeutic index of levodopa. The patients are benefited by lowering the dose of levodopa or giving levodopa with selegiline (selective MAO-B inhibitor).

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Section 5

GENITOURINARY SYSTEM

The cardinal symptoms of (genito-) urinary system are :

1. Swelling of the face, legs or whole body (i.e., anasarca)
2. Anuria
3. Oliguria
4. Polyuria
5. Haematuria (bloody urine)
6. Nocturia (all polyuric states, oedema-forming states, prostatism, salt-losing nephropathy, cystitis, vesico-ureteric reflux, and insomnia even without renal disease produce nocturia). In CCF and other oedema-forming states, there is increase in renal blood flow during recumbency.
7. Dysuria (pain or discomfort during micturition), strangury, burning micturition, frequency of micturition (passing urine more often than usual) and urgency (sudden urge or need to pass urine); hesitancy (delay in initiation) or precipitancy (sudden voiding of urine), post-micturition dribbling (e.g., benign hypertrophy of prostate—BHP), impaired flow or urine, or urethral discharge
8. Retention of urine
9. Incontinence of urine (stress, urge, true or overflow incontinence)—vide page 171
10. Enuresis (bed-wetting at night or during sleep)
11. Increased nocturnal frequency (BHP, diabetes mellitus, nocturia)
12. Fork urine (bifid) in stricture urethra; pneumaturia (bubbly urine) in vesico-enteric fistula (e.g., in Crohn's disease) or in gas producing organisms in the urinary tract in diabetes mellitus.
13. Passage of milkly urine—Chyluria; smoky urine (AGN); foul smell in urine (UTI)
14. Swelling of abdomen (ascites, renal lump)
15. Loin pain
16. Renal colic
17. Sexual problems like lack of libido, impotence, trouble with ejaculation; dyspareunia and menstrual problems in female. Any symptom related to sexually transmitted diseases (STD).
18. Any complaint regarding male or female genitalia
19. Fever (UTI, renal tuberculosis, pyelonephritis)
20. General symptoms—nausea/vomiting/hiccough (renal failure), appetite, weight loss/gain (i.e., oedema), sleep, fatigue, bowel etc.
21. Miscellaneous—pruritus, breathlessness, paraesthesia. restless legs are features in renal failure.

* BHP is manifested by nocturia, increased frequency, reduced size and force of urinary stream, straining to urinate and dribbling at the end.

** **Dysuria** is difficulty in passing urine which may be associated with pain or discomfort. **Strangury** is painful (severe) passing of urine drop by drop.

Scheme of Examination

This system may be written separately though few clinicians prefer the combined system of G.I. tract plus genitourinary system. While writing this system, proforma of G.I. system should be followed with special reference to :

(A) GENERAL SURVEY :

1. Pallor with yellow complexion (e.g., CRF).
2. Face—Puffiness of the face or moon face, present or not.
3. **Oedema**—Pitting or non-pitting, localised or generalised, parietal oedema present or not.
4. Nutrition—Cannot be assessed properly due to presence of anasarca (if present).
5. Pulse—Condition of the arterial wall may be thickened rarely in nephrotic syndrome (due to accelerated atherosclerosis). Sometimes, an arteriovenous fistula (Brescia-Cimino fistula; vascular access artificially made for haemodialysis) with a continuous thrill may be present in the anterior aspect of lower part of forearm.
6. BP measurement is a must in all patients.
7. Skin—Whether scabies with **pyoderir** a present (acute nephritis) or not.

(B) G.I. SYSTEM : In details (**tonsils** should be examined in a patient with acute nephritis; **prostate** should be examined per rectally in suspected obstructive uropathy).

(C) GENITOURINARY SYSTEM :

- I. Inspection :
Genitalia—Penile swelling, vulval oedema, scrotal swelling, contact ulcer etc. Observe the skin for renal biopsy mark (benzene seal or leucoplast strap in the loin), nephrectomy scar (in the loin), renal transplant scar (in either of iliac fossa), and small scar or catheter in situ in the centre of lower abdomen for peritoneal dialysis.
*** Testis :**
Small—hypogonadism, absence—cryptorchidism, tenderness—orchitis, tumour teratoma, seminoma; also look for hydrocele and inguinal hernia.
**** Penis :**
Adult size is approximately 8-12 cm in length. In hypogonadism, the size is < 2.5 cm.
- II. Palpation :
 1. Examination of genitalia for **phimosis**, scrotal oedema, hydrocele, contact ulcer in genitalia, palpation of **testes** etc.
 2. Parietal oedema.
 3. Fluid thrill.
 4. Detailed examination of kidneys (palpation) with special reference to bimanual palpation and ballottement to find out a **renal mass**.
 5. **Renal angle**—Tender or not; fullness of renal angle in perinephric abscess.
- III. Percussion :
 1. Shifting dullness in abdomen.
 2. Upper border of liver dullness (as hydrothorax may be present).
 3. Band of colonic resonance over the renal mass.
 4. Percussion of the urinary bladder.
- IV. Auscultation :
 1. Peristalsis.
 2. Renal artery bruit (heard on either side of the midline in the mid-abdomen in renal artery stenosis).
 3. Venous hum.
- V. In detail examination of genitalia, hernial orifices, per rectal or per vaginal examination (if indicated).

(D) CVS:

1. Neck veins may not be properly visualised due to anasarca, or neck veins remain engorged due to fluid overload or pericardial tamponade.
2. Apex—Site (for LVH arising out of renal hypertension), murmur, gallop etc.
3. **Pericardial rub** (uraemia).
4. Pericardial effusion (percussion of the heart may be necessary).

* CCF due to renal diseases are common as a result of fluid overload and hypertension.

(E) RESPIRATORY SYSTEM :

1. Hyperpnoea due to metabolic acidosis in renal failure.
2. Parietal oedema in the chest wall (i.e, intercostal fullness).
3. Type of respiration-May be thoraco-abdominal due to the presence of ascites
4. Features of **hydrothorax**.
5. Features of lower respiratory tract infection.

(F) NERVOUS SYSTEM :

(G) •URINE EXAMINATION (OPTIONAL, : For the presence of albumin and sugar

foreskin, may be responsible, or "S" TM, 'p5, ritu P,

II*

look for bilateral mark in loin (for kidney) in a patient with nephrotic syndrome.
 in history taking, past history (diabetes, hypertension, anaemia, recurrent infection) family history

SV,,?or,'s syndrome"and drug histA

Case 20

ACUTE GLOMERULONEPHRITIS

What is your diagnosis ?

It is a case of acute glomerulonephritis probably poststreptococcal in aetiology.

Why do you say so ?

This is a case of acute glomerulonephritis (AGN) because :

1. This boy aged 12 years presented with swelling of his face, scanty micturition with haematuria (r smoky urine) for last four days. The disease was acute in onset and progressive in nature. He was absolutely normal prior to four days. His mother first noticed the puffiness of her son's face (mainly periorbital) in the morning after getting up from the bed. He complained of oliguria and haematuria from the same day. There was no H/O respiratory distress convulsions or anuria. He did not complain of any burning micturition or loin pain'(due to stretching of renal capsule). There was presence of low grade fever at the initial period with anorexia, headache and malaise but H/O vomiting was absent.
2. Though he did not suffer from any sore throat (e.g., tonsillitis, pharyngitis) 1-3 weeks back he gave H/O generalised itching all over the body for last 3 weeks which was probably due to cables infection. There was neither any H/O viral hepatitis B, chickenpox, mumps measles infection in the recent past nor there was any past history suggesting collagen vascular disease drug hypersensitivity, type I diabetes or hypertension. disease,
3. On examination the face is pale and puffy (**nephritic facies**). The swelling is seen more in the eyelids (periorbital) causing narrowing of the palpebral fissure. There is presence of mild pitting, pedal oedema at present. Pyoderma (infected scabies) is seen in the classical distribution like webs of fingers, elbow, below the nipple, umbilicus and groin regions. Pulse shows tachycardia and the BP is 150/90 mm of Hg (definitely hypertensive at the age of 12 years). Jugular veins are engorged (due to hypervolaemia).
4. Neither scrotal oedema nor ascites is detected. Liver, spleen and kidneys are not palpable and the renal angles are non-tender. p p 'la
5. Other systems are within normal limit.
6. Urine examination done at the bedside revealed presence of albumin but there is absence of

con-oboration°

b'°°d

"mP'e * US"a"y n°i

be P^rtomed for

* Sometimes, a patient of AGN may present with anasarca. The 'latent period' in case of pharyngeal infection is usually 1-3 weeks and in cutaneous infection (pyoderma from scabies, furunculosis or impetigo), it is averaging about 2-6 weeks. p

What is your case ?

Say the summary as mentioned above.

What is acute glomerulonephritis (AGN) or acute nephritic syndrome ?

It consists of sudden onset symptom complex like :

1. Periorbital puffiness.
2. Oliguria.
3. Haematuria.
4. Oedema and hypertension.
5. Proteinuria (non-nephrotic range).
6. RBC and RBC cast in urine.
7. Tendency of early renal failure (acute).

Possible pathogenesis of symptomatology in AGN :

1. Oedema (periorbital, legs or sacral)
 - Retention of salt and water, and
 - Hypoproteinaemia.
2. Hypertension—
 - Stimulation of renin-angiotensin-aldosterone axis, and
 - Hypervolaemia due to salt and water retention.
3. Haematuria (microscopic)—
 - Glomerular inflammation
4. Periorbital swelling or puffiness—
 - Collection of oedema fluid in loose periorbital tissue.
5. Oliguria—
 - Reduction in glomerular filtration rate (GFR).
6. Uraemia or renal failure—
 - Retention of urea, creatinine and other nitrogenous waste products.

Relevant 'past history' in acute nephritis :

1. Sore throat (streptococcal). \ poststreptococcal glomerulonephritis (PSGN)
3. Infections like—Heptims[^], mumps, measles, chickenpox, infectious mononucleosis, malaria, pneumococcal pneumonia, infective endocarditis, syphilis (secondary).
4. Collagen vascular diseases (SLE, vasculitis, Henoch-Schonlein purpura etc.)
5. Drug hypersensitivity, G.B. syndrome.
6. 'Shunt nephritis' (infection of ventriculo-atrial shunt done in hydrocephalus).
7. H/O diabetes and hypertension.
8. H/O prolonged steroid therapy (ID/D of moon face) or urticaria (angioneurotic oedema as a D/D of moon face).

* Actually 1 to 6 are the different causes of AGN and poststreptococcal aetiology is the commonest among them.

Physical findings in other systems in acute nephritis :**(A) CVS:**

- (i) JVP may be raised; systemic hypertension.
- (ii) Apex may shift down and out (due to LV dilatation), and forceful in nature.
- (iii) **loud A**, (due to hypertension).
 - (iv) There may be presence of mitral systolic murmur (functional murmur due to LV dilatation).
- (v) Gallop rhythm may be present (LVF).
- (vi) Rarely, pericardial rub in acute renal failure.

(Li) RESPIRATORY SYSTEM :

- (i) Bilateral basal crepitations (due to LVF).
- (ii) Usually, there is absence of hydrothorax (to differentiate it from nephrotic syndrome).

(C) NERVOUS SYSTEM :

- (i) Features of hypertensive encephalopathy.
- (ii) Fundoscopy—Usually normal (not done).

(D) *G.I. TRACT* : Tonsils may be enlarged with tender cervical adenopathy due to acute streptococcal tonsillitis. Ascites may be vident.

(E) *SKIN* (in general survey) : Scabies with pyoderma.

* Never forget to examine the patient for phimosis.

Common features in adults with AGN :

1. There may not be any H/O preceding infection.
2. Oedema is not marked.
3. Hypertension may or may not be present.
4. Presence of oliguria, haematuria and proteinuria (as occurs in a child).

Pathogenesis and types of streptococci involved in AGN :

Poststreptococcal AGN is produced due to infection by group A beta-haemolytic streptococci (nephritogenic strains). *Remember, streptococcal pharyngitis may produce AGN or acute rheumatic fever while streptococcal skin infection produces AGN only.*

M types of streptococci (nephritogenic strains), e.g., types 2, 47, 49, 55, 57 and 60 are seen following skin infections, and types 1, 2, 3, 4, 12, 25 and 49 after pharyngitis (throat infection).

Actually, AGN or acute nephritic syndrome is an immune complex glomerulonephritis where there is production of antibodies against glomerular antigen and thereby leads to deposition of immune complexes in the walls of glomerular capillaries, which ultimately develops into inflammation of the glomeruli.

Causes of acute glomerulonephritis :

- I. Infectious diseases :
 - (A) Poststreptococcal glomerulonephritis.
 - (B) Non-poststreptococcal glomerulonephritis.
 - a) Bacterial—staphylococcal and pneumococcal infection, infective endocarditis, meningococcaemia, typhoid.
 - b) Viral—hepatitis B, mumps, measles, varicella, infectious mononucleosis, coxsackievirus.
 - c) Parasitic—malaria.
- II. Systemic disorders :

Vasculitis, SLE, Henoch-Schonlein purpura, Goodpasture's syndrome.
- III. Primary glomerular disease : membranoproliferative glomerulonephritis, mesangial proliferative glomerulonephritis, IgA nephropathy (Berger's disease).
- IV. Miscellaneous : G.B. syndrome, serum sickness, DPT vaccination.

Causes of red urine in clinical practice :

1. Haematuria (presence of intact RBC under microscope).
 2. Haemoglobinuria (in haemolytic disorders; urine gives chemical tests for haemoglobin but there is no RBC present in the centrifuged deposit of fresh sample of urine) and myoglobinuria (in rhabdomyolysis; very dark urine; no RBC on M/E)*.
 3. Ingestion of beet root, senna and dyes used to colour sweets.
 4. Phenolphthalein (used in purgatives) in alkaline medium (after use of systemic alkalisers) of urine.
 5. Acute intermittent porphyria (presence of porphobilinogen may be detected by addition of Ehrlich's aldehyde reagent. Freshly voided urine appears normal but may take dark red colour on standing for few hours).
 6. Drugs—Phenazopyridine (pyridium), phenindione, clofazimine, rifampicin (it actually changes the urine to dark orange colour).
 7. Alkaptonuria (becomes black, if kept for long time).
- * If blood from these patients are kept in a test-tube for few minutes, the supernatant serum will be reddish in myoglobinuria while it will be clear in patients with haemoglobinuria.

Medical causes of haematuria :

- | | |
|---|---|
| 1. Acute glomerulonephritis. | 7. Anticoagulant therapy. |
| 2. SBE (generally microscopic haematuria). | 8. Renal tuberculosis. |
| 3. Malignant hypertension. | 9. Papillary necrosis (diabetes, sickle cell disease, analgesic-induced). |
| 4. Snake bite. | 10. SLE (microscopic haematuria). |
| 5. Henoch-Schonlein purpura or other coagulation disorders. | 11. Goodpasture's syndrome. |
| 6. Weil's disease. | |

* **Painless haematuria** : AGN, SBE, IgA nephropathy, hypernephroma, coagulation disorder, renal tuberculosis, hypertension. **Painful haematuria** : urinary tract infection, renal calculus and trauma.

Definition of polyuria, oliguria and anuria :

1. **Polyuria**—Urine output persistently above 3 litres per day. The normal urine output for a healthy adult is approximately 1.5 litre (400 ml- 3 litre) per day. The common causes are :
 - a) Chronic renal failure.
 - b) Diabetes mellitus.
 - c) Diabetes insipidus.
 - d) Psychogenic polydipsia (compulsive water drinking).
 - e) Use of diuretics in patients with oedema.
 - f) Diuretic phase of ARF.
 - g) Hypercalcaemic nephropathy.
 - h) Hypokalaemic nephropathy.
 - i) 'Post-obstructive uropathy'.
 - j) Drugs—alcohol, caffeine.
 - k) Transient polyuria (after epileptic seizure, paroxysmal atrial tachycardia)
2. **Oliguria**—Urine volume < 400 ml per day. Common causes are acute gastroenteritis, high fever, acute glomerulonephritis, congestive heart failure (decompensated), renal failure (acute and chronic), hypovolaemia and shock, 'third space' loss in acute peritonitis and pancreatitis.
3. **Anuria**—No urine formation for 12 hours (some nephrologists define anuria as < 50 ml urine output per day). Complete anuria, i.e., no urine formation by catheterisation occurs in bilateral ureteric obstruction, rapidly progressive glomerulonephritis, diffuse cortical necrosis and bilateral renal artery stenosis.

* Passing of more than one-third of the total daily output by night is **nocturia**, and diabetes mellitus and benign hypertrophy of prostate are two prime causes in clinical practice.

Why this is not a case of nephrotic syndrome ?

As there is :

1. History of scabies with secondary infection.
2. 'Acute onset' of oliguria.
3. Absence of anasarca.
4. Presence of haematuria.
5. Presence of hypertension.
6. A classical history.
7. Microscopical examination of urine (if allowed to do) shows presence of mild to moderate proteinuria, RBC and RBC cast.

Differential diagnosis of your case :

1. Angioneurotic oedema—Itchy swelling of eyelids, lips and tongue; absence of pyrexia, hypertension, oliguria and haematuria. H/O atopy present.
2. Acute pyelonephritis—Fever with chill and rigor; H/O loin pain with tender renal angles. Oedema is absent and there is no hypertension. Increased frequency of micturition is often present.
3. Other causes of haematuria are to be differentiated.
4. Nephrotic syndrome—Already mentioned above.
5. Acute exacerbation of chronic nephritis—H/O previous attacks of nephritis, severe hypertension and renal functional impairment are present.

Investigations you like to do :

1. Urine examination—
 - (i) Smoky urine.
 - (ii) 300-600 ml In 24 hours.
 - (iii) High specific gravity (due to increased tubular reabsorption).
 - (iv) **Moderate albuminuria.**

- (v) Presence of RBC, RBC cast, WBC, granular cast (present lately), epithelial cell and cast **(RBC cast is diagnostic of acute nephritis).**
 - (vi) Urine is sterile on culture.
 - 2. Blood—
 - (i) Anaemia (haemodilution).
 - (ii) Leucocytosis.
 - (iii) Raised ESR.
 - (iv) Raised ASO titre in poststreptococcal AGN (more common with throat infection).
 - (v) Hypocomplementaemia C₃ and C₄.
 - 3. Blood biochemistry—
 - (i) Urea and creatinine levels may be elevated (if progresses to acute renal failure).
 - (ii) Normal cholesterol (differentiates nephrotic syndrome).
 - 4. Chest X-ray—
 - (i) Cardiac enlargement, or
 - (ii) Features of pulmonary congestion (indicates hypervolaemia).
 - 5. Renal function test—Diminished GFR.
 - 6. Cultures of throat swab or swab from infected skin may grow group A beta-haemolytic streptococci.
 - 7. Ultrasonography of kidneys shows normal or large kidneys.
- * **Characteristic cast in AGN is RBC cast, in nephrotic syndrome it is fatty cast, granular cast in CRF, and WBC cast in chronic pyelonephritis.**
- ** Azotaemia' is the increase in the concentration of urea and creatinine in blood resulting from a fall in the glomerular filtration rate (GFR).
- *** Renal biopsy is usually not done in acute glomerulonephritis.

Complications of acute nephritis :

- 1. Acute renal failure (ARF).
- 2. Hypertensive encephalopathy.
- 3. Acute left ventricular failure (LVF).
- 4. Infections—Urinary tract, respiratory tract etc.
- 5. Fluid and electrolyte imbalance, nephrotic syndrome, chronic glomerulonephritis.
- 6. Fluid overload.
- 7. Rapidly progressive glomerulonephritis (RPGN).

How to recognise the major complications of AGN ?

- (A) ACUTE RENAL FAILURE :
 - (i) Severe oliguria to anuria.
 - (ii) Drowsiness, confusion, delirium and even coma may supervene.
 - (iii) Hypertension.
 - (iv) Acidotic breathing (deep sighing, rapid breathing at a regular rate and with a hissing sound is called Kussmaul's breathing).
 - (v) Blood urea, creatinine and potassium levels are invariably high with acidosis.
- (B) HYPERTENSIVE ENCEPHALOPATHY :
 - (i) Features of increased intracranial tension, i.e., headache, vomiting, convulsions or coma.
 - (ii) Hypertension.
 - (iii) Confusion, delirium.
 - (iv) Though rare, focal neurological signs like temporary blindness, aphasia, hemiparesis or extensor plantar response may be present.
 - (v) Papilloedema with retinal exudates and haemorrhages may be evident.
- (C) ACUTE LEFT VENTRICULAR FAILURE :
 - (i) Acute respiratory distress, orthopnoea; H/O PND.
 - (ii) Cough with copious, pinkish, frothy sputum.
 - (iii) Central cyanosis.

- (iv) Hypertension.
- (v) Gallop rhythm, crepitations at lung bases and pulsus alternans.

* **These 3 are the major complications of AGN.** Prognosis of (A), (B) and (C) are very bad. All the complications should be treated promptly as soon as they are recognised.

What is RPGN?

It is known as rapidly progressive glomerulonephritis (RPGN). This term is often applied to a group of acute nephritis patients who develop persistent renal failure in a period of weeks to months and show little tendency for spontaneous recovery. Usually the pathology is extensive extracapillary (crescentic) glomerulonephritis. Clinically there are presence of nausea, vomiting, weakness, persistent hypertension, haematuria, proteinuria and azotaemia. Common causes are systemic vasculitis, SLE, poststreptococcal AGN, Goodpasture's syndrome. The prognosis is poor. Steroids, cytotoxic agents or intensive plasma exchange may be useful. Fifty percent of patients may require dialysis within 6 months.

Common causes of acute renal failure (ARF) in your hospital :

- | | |
|---|-------------------------------------|
| 1. Hypovolaemia due to acute gastroenteritis, severe haemorrhage, pancreatitis, peritonitis or extensive burns. | 5. Mismatched blood transfusion |
| 2. Acute glomerulonephritis. | 6. Snake bite. |
| 3. Septicaemia, endotoxic shock. | 7. Septic abortion, eclampsia. |
| 4. Malignant hypertension. | 8. Acute myocardial infarction. |
| | 9. Hepato-renal syndrome. |
| | 10. Acute interstitial nephropathy. |

Prognosis of acute nephritis :

1. Complete recovery—85% of children and 50% of adults (within 2 weeks).
2. Complications—5% cases produce complications mentioned above; few suffer from RPGN.
3. Apparent recovery—10% cases live with persistent proteinuria and haematuria for long periods.

* As a whole, the prognosis is excellent and the prognosis in children is better than adults.

How do you like to manage a case of AGN?

1. Strict bed rest—Continued until the signs of glomerular inflammation and circulatory congestion subside (i.e., disappearance of oedema, oliguria, haematuria, hypertension, high blood urea, RBC cast in urine).
2. Diet—Mild protein restriction is done in azotaemic patients. When oliguria is moderate, 0.5 mg/kg/day of protein should be given. Mainly carbohydrate diet is preferred.
3. Salt and water balance—Salt-free foods are given. The fluid intake is restricted to previous days urine output plus 500 ml of fluid. Salt and fluid restriction are most important.
4. Diuretics—Tablet frusemide (40 mg/tab) 1 tab daily is given to relieve oedema and hypertension.
5. Antihypertensives—nifedipine, amlodipine, hydralazine and diazoxide may be of help.
6. Antibiotics—7 to 10 days course of crystalline penicillin (5 lakhs, I.M, BD) or erythromycin (250 mg QDS, orally) is given if streptococcal infection is documented.
7. Treatment of complications.
8. Dialysis may be required in severely azotaemic or fluid overloaded patients.
9. Steroid, cytotoxic drugs, long-term chemoprophylaxis like rheumatic fever are not needed.
10. Tonsillectomy may be done afterwards.

Conclusion :

1. Always measure the BP of the patient.
2. Examine for phimosis, scabies infection and swelling of tonsils.
3. Percuss the urinary bladder for retention of urine (to make D/D with oligo-anuria).
4. Search for gallop rhythm and pericardial rub.
5. Note the colour of the urine carefully (red, smoky or coca-cola urine)—send the patient to toilet with a test tube to collect urine.

N.B. : Acute glomerulonephritis is also known as acute nephritis. Type I nephritis, acute glomerulonephritis syndrome or acute nephritic syndrome.

Case 21

NEPHROTIC SYNDROME

What is your diagnosis ?

The diagnosis may be delivered in one of the two ways :

1. This is a patient of nephrotic syndrome probably due to minimal lesion nephropathy, or
2. This is a patient of anasarca probably due to nephrotic syndrome.

Why do you say so ?

This is because of :

1. This male child aged 14 years and 2 months presented with gradual swelling of the whole body for last one month. He was absolutely normal one month back. The disease was insidious in onset and gradually progressive. His father first noticed the puffiness of his son's face (mainly periorbital) one morning where the swelling gradually involved the whole body (descending oedema) in course of time. The swelling was a bit posture-dependent. Lately, swelling involved the abdomen and the scrotum; it was painless and pitting in nature according to the patient. The boy also complained of increasing weakness. The output and colour of urine were within normal limit, and there was no H/O haematuria, burning micturition, renal colic. He did not give any H/O respiratory distress.
 2. There was no past H/O sore throat, skin infection, diabetes mellitus, drug intake or jaundice; no family H/O diabetes or hypertension was present, and no other family member was suffering from the same type of illness.
 3. On examination, the face is puffy with baggy lower eyelids and It seems a 'moon face' (also there is waxy pallor in the face). There is presence of generalised, massive, posture-dependent, descending pitting oedema. BP is within normal limit (110/80 mm of Hg). Scrotal oedema with contact ulcers, and parietal oedema are present. Huge swelling of penis is seen. There is mild anaemia.
 - 4 Fluid thrill and shifting dullness are present, i.e.. there is presence of ascites. Liver, spleen, gall bladder and kidneys are not palpable. Renal angles are non-tender.
 5. On examination of the other systems there is no pericardial rub, hydrothorax or pericardial effusion present.
 5. Urine examination done at the bedside revealed presence of albumin but there is absence of sugar in the sample—may be performed for corroboration.
- * Renal oedema : periorbital + genital + generalised oedema.

Causes of anasarca and moon face :

Read the sections on 'Oedema' and 'Moon face' respectively.

What is nephrotic Syndrome ?

It is a clinical syndrome characterised by :

- a) Massive proteinuria (albuminuria).
- b) Hypoalbuminaemia (hypoproteinaemia),
- c) Generalised oedema or anasarca, and
- d) Hyperlipidaemia (e.g., hypercholesterolaemia).

* Proteinuria > 3.5 g/ 1.73 m² per day is considered to be in the nephrotic range. Normal body surface for a reference man is 1.73 m². *Proteinuria is the hallmark of nephrotic syndrome.*

** Now-a-days, lipiduria and hypercoagulability are new additions in the clinical complex of nephrotic syndrome.

Importance of 'past history' in nephrotic syndrome :

(A) Relating to aetiology :

1. Jaundice (hepatitis B infection).
2. Sore throat (poststreptococcal).
3. Diabetes mellitus.
4. Diseases like leprosy, syphilis, malaria.
5. Drug Intake—Anti-snake venom, penicillamine, troxidone, probenecid, organic gold.
6. Swelling in the neck (lymphoma).
7. Skin rash, arthralgia, arthritis, alopecia (collagen vascular disease).

(B) Relating to cause of anasarca :

1. Jaundice, alcoholism, H/O haematemesis or melaena (indicates cirrhosis of liver).
2. H/O respiratory distress or palpitation (cardiac disorder).
3. H/O chronic diarrhoea (malabsorption and malnutrition).
4. H/O tuberculosis (tuberculous pericardial effusion or constrictive pericarditis).
5. Hoarseness of voice and constipation (hypothyroid).
6. H/O long-standing corticosteroid therapy (D/D with moon face and swelling of body).

How nephrotic syndrome is classified ?

1. Primary glomerular diseases :
 - a) Minimal lesion or minimal change disease.
 - b) Membranous glomerulonephritis.
 - c) Focal and segmental glomerulosclerosis.
 - d) Membranoproliferative glomerulonephritis (or mesangiocapillary glomerulonephritis).
 - e) Mesangial proliferative glomerulonephritis.
2. Secondary to other diseases :
 - a) Infections—Poststreptococcal, hepatitis B, malaria, lepromatous leprosy, secondary syphilis, infectious mononucleosis, infective endocarditis, HIV infection and AIDS.
 - b) Drugs—Anti-snake venom, penicillamine, troxidone, probenecid, bismuth, gold, mercury, captopril, α -interferon, street heroin, contrast media.
 - c) Multisystem disease—SLE, Henoch-Schonlein purpura, vasculitis due to any cause, rheumatoid arthritis, Goodpasture's syndrome.
 - d) Neoplasm—Lymphoma, leukaemia, carcinoma, melanoma.
 - e) Heredo-familial—Diabetes mellitus, congenital nephrotic syndrome, Alport's syndrome, sickle cell disease, nail-patella syndrome.
 - fj Miscellaneous—Pre-eclamptic toxemia, amyloidosis, snake bite, bee-stings.

What is Kimmelstiel-Wilson (K-W) syndrome ?

1. It is a variety of nephrotic syndrome commonly arising from type I diabetes mellitus (30-40% in Type I and 3-10% in type II diabetes mellitus).
2. Histological lesion is nodular glomerulosclerosis.
3. Usually develops in a long standing diabetes mellitus (>10 years approximately).
4. Commonly starts with proteinuria.
5. The requirement of insulin becomes less with the development of K-W syndrome (as with development of nephropathy, insulin antibodies come out with urine).
6. Lately hypertension may be present.
7. End-stage renal disease can be expected to occur within 5-7 years.

* K-W lesion : Nodular glomerulosclerosis (K-W nodules, reasonably specific for diabetes mellitus).

K-W disease : Any manifestation of glomerulonephropathy in diabetes mellitus.

K-W syndrome : Nephrotic syndrome from diabetes mellitus.

Common causes of nephrotic syndrome in different age group :**(A) Children—**

1. Minimal lesion (70%).
2. Mesangial proliferative (15%).
3. Focal glomerulosclerosis (10%).
4. Secondary—5%.
5. Membranous, membranoproliferative—Rare.

children, primary disease is 95% and secondary is 5%:
secondary is 30%.

(B) Adults—

1. Membranous—35 %.
2. Secondary—30 %.
3. Minimal lesion—15%.
4. Focal glomerulosclerosis—10%.
5. Mesangial proliferative—5%.
6. Membranoproliferative—<5 %.

; In adults, primary disease is 70% and

Possible pathogenesis of different features in nephrotic syndrome :

- Proteinuria—Either due to change in pore size or J- in anion barrier in lining of foot process of the epithelium.
- Hypoalbuminaemia—Proteinuria plus renal catabolism of filtered albumin.

- Oedema and anasarca—Hypoalbuminaemia in oncotic pressure \rightarrow T in extracellular fluid volume. Moreover, there is **I** in intracellular fluid \rightarrow T renin-angiotensin-aldosterone axis \rightarrow T Na and water retention \rightarrow oedema and anasarca.
- Hyperlipidaemia and lipiduria—Hypoalbuminaemia \rightarrow **I** oncotic pressure \rightarrow stimulate liver for lipoprotein synthesis \rightarrow T cholesterol, t LDL, T TG and T VLDL ^hyperlipidaemia may be responsible for accelerated atherosclerosis and progression of renal disease.
- Hypercoagulability— I antithrombin III, protein C and S, **i** fibrinolysis, hyperfibrinogenaemia and t platelet aggregation \rightarrow T coagulability of blood with thromboembolic manifestations.

Differential diagnosis of nephrotic syndrome :

(A) Causes of anasarca :

1. CCF—Read the section on 'Oedema'.
2. Cirrhosis of liver—
 - (i) H/O haematemesis and melaena may be present.
 - (ii) Usually the face is not puffy.
 - (iii) Ascites appears first; pedal oedema appears next.
 - (iv) Splenomegaly and venous prominence in the abdomen with venous flow away from the umbilicus.
 - (v) Signs of hepato-cellular failure may be present.
3. Anaemia with hypoproteinaemia—
 - (i) Pallor.
 - (ii) Signs of malnutrition like glossitis, angular stomatitis, cheilosis, koilonychia may be present.
 - (iii) Oedema appears first in ankles: ascites appears late.
 - (iv) Systolic murmur may be present over the pulmonary area.
4. Constrictive pericarditis—Read the section on 'Examination of neck veins'.
5. Pericardial effusion—Read the section on 'Pericardial rub'.
6. Myxoedema—
 - (i) Absence of urinary problem.
 - (ii) Hoarseness, constipation, cold sensitivity, lethargy, menorrhagia.
 - (iii) Bradycardia, hypertension.
 - (iv) Oedema is non-pitting in nature.
 - (v) Delayed relaxation of ankle jerk.

(B) Causes of moon face : Read the sections on 'Moon face' and 'Acute glomerulonephritis'.

Common complications of nephrotic syndrome :

1. Protein catabolism—May lead to wasting, striae and osteoporosis.
2. Impaired resistance to infection like cellulitis, respiratory tract infection, peritonitis.
3. Thromboembolic manifestations (due to loss of antithrombin III in urine, hyperfibrinogenaemia, impaired fibrinolysis, reduced activity of protein C or protein S, hyperlipidaemia, and enhanced platelet aggregation)—Arterial thrombosis (pulmonary artery thrombosis specially), venous thrombosis (renal vein thrombosis specially).
4. Hypovolaemia may produce syncope, postural hypotension, renal failure.
5. Renal failure (chronic).
6. Accelerated atherosclerosis (may lead to 'strokes in young and early coronary artery disease).
7. Fluid retention, hydrothorax, pericardial effusion.
8. Consequences of loss of specific binding proteins (see below).

Renal vein thrombosis in nephrotic syndrome :

Previously it was thought to be a cause of nephrotic syndrome but now-a-days it is regarded as a consequence. The condition is treated by long-term oral anticoagulation. It is commonly found in :

1. Membranous nephropathy,
2. Membranoproliferative glomerulonephritis, and
3. Amyloidosis.

It is diagnosed by :

- | | |
|---------------------------------------|---|
| a) Unilateral or bilateral loin pain. | d) Widely fluctuating urine volume. |
| b) Haematuria. | e) Asymmetry of renal size in ultrasonography. |
| c) Left-sided varicocele. | f) Best documented by renal venous angiography. |

Nephrotic syndrome with hypertension :

Usually hypertension is not a feature of nephrotic syndrome. If present, one should think of :

1. K-W syndrome (i.e., diabetic nephropathy).
2. SLE as an aetiology.
3. Polyarteritis nodosa as an aetiology.
4. The patient is on long-term corticosteroid therapy.
5. Nephritic-nephrotic syndrome—nephrotic syndrome with atypical features of hypertension, microscopic or gross haematuria, early renal failure and low complement C₃. The common aetiologies are SLE and Henoch-Schonlein purpura.
6. The patient has developed early CRF.

* Focal glomerulosclerosis is commonly associated with hypertension,

i** Interstitial nephritis usually have normal blood pressure].

Physical signs in different systems in nephrotic syndrome :

1. **G.I. tract and renal system**—Skin of the abdomen is shiny, ascites, neither hepatosplenomegaly nor any renal lump felt, non-tender renal angle, scrotal (or vulval) oedema or presence of hydrocele; urinary bladder is generally empty. Phimosis should be looked for.
2. **Respiratory system** Respiration is thoraco-abdominal in type (due to presence of ascites), parietal oedema in the chest wall, bilateral hydrothorax, crepitations at lung bases may be audible. Sometimes, evidences of pneumonia is present.
3. **CVS**—The patient may be orthopnoeic. BP is usually normal. Neck veins may not be properly visualised due to oedema (usually JVP is not raised); presence of pericardial rub (in renal failure), rarely pericardial effusion; condition of the arterial wall (in peripheral pulse) may reveal thickening of arteries due to accelerated atherosclerosis (press the oedema fluid for few seconds for better palpation of peripheral pulse).
4. **Nervous system**—Nothing particular (patient may be lethargic or drowsy in the presence of renal failure).
5. **Reticulo-endothelial system**—Examine the patient for lymphadenopathy (as lymphoma, SLE and infectious mononucleosis may be the aetiology). Absence of sternal tenderness.

* Xanthelasma around the eyes may be occasionally seen because of hypercholesterolaemia.

In nephrotic syndrome, consequences of plasma proteins lost in urine :

1. Albumin (oedema, anasarca and increased susceptibility to infections).
2. Antithrombin III (causes increased coagulability and thromboembolic complications).
3. Thyroxine-binding globulin (produces abnormal thyroid function tests).
4. Cholecalciferol-binding protein (osteomalacia may be precipitated).
5. Transferrin (produces iron-resistant microcytic-hypochromic anaemia).
6. Metal binding proteins (may produce Zn and Cu-deficiency states).
7. Globulin (severe IgG deficiency may lead to bacterial peritonitis).
8. Drug-binding proteins (may result in altered drug pharmacokinetics).

How do you classify proteinuria ?

Normal adults may excrete upto 150 mg protein in 24 hours of which 15 mg is albumin and 25 mg is Tamm-Horsfall mucoprotein' produced by the renal tubules. Proteinuria (pathological) is classified into :

- a) Mild—150-500 mg/day (+)
- b) Moderate—500 mg-2g/day (++)
- c) Massive >2g/day (+++); if crosses 3.5 g/day, it is designated as 'nephrotic range'.

* Proteinuria may be divided into selective (responsive to corticosteroid) and non-selective types.

** **Microalbuminuria** : It is defined as daily excretion of albumin of 20-200 microgram/min (i.e. 30-300 mg/day) and is commonly found in diabetes mellitus with early renal involvement.

*** Proteinuria often makes the urine frothy.

Characteristics of minimal change disease :

1. It is also known as lipoid nephrosis, nil lesion or foot process disease. No alterations are demonstrable by light microscopy (so, nil lesion) but electron microscopy shows diffuse epithelial foot process effacement.

2. It is the commonest cause of nephrotic syndrome in children (70%); male > female.
3. Highly selective proteinuria.
4. Progression to acute renal failure is rare and the prognosis is good as 10 years survival rate exceeds 90%.
5. Generally steroid or cytotoxic drug responsive.

Characteristics of membranous nephropathy :

1. It is the commonest form of nephrotic syndrome in adults (35%).
2. Commonly due to SLE, carcinoma of lung or penicillamine therapy.
3. Light microscopy shows thickening of glomerular basement membrane (GBM)—so called ‘membranous’.
4. Non-selective proteinuria. Haematuria may be evident.
 5. Renal vein thrombosis is common, as common is the progression to chronic renal failure.
6. Prognosis is bad.
7. Treated by steroid but the response is inconsistent.

Neuromuscular complications of chronic renal failure (azotaemia) :

1. Fatigue 2. Headache 3. Lethargy 4. Difficulty in concentration 5. Drowsiness 6. Muscular irritability 7. Flapping tremor (asterixis) 8. Hallucinations 9. Convulsions 10. Peripheral neuropathy 11. Myoclonus 12. Muscle cramps 13. Restless leg syndrome 14. Coma.

How do you like to investigate your case ?

- (A) Urine examination—Urine volume is normal or low, **massive proteinuria** (24-hours urinary protein estimation is essential for diagnosis), normal specific gravity, shows hyaline and **fatty cast**.
- (B) Blood examination—
 - (i) Mild normocytic normochromic anaemia.
 - (ii) Albumin level < 3 g/dl (normal level of albumin is 3.5 to 5.5 g/dl and that of globulin is 2 to 3.5 g/dl). Often the α_2 and β globulin are elevated inspite of low total globulin level.
 - (iii) Urea and creatinine levels are usually normal but may be elevated in renal failure.
 - (iv) Serum cholesterol is high and is usually > 300 mg/dl (normal desirable value <200 mg/dl).
 - (v) Others—Blood sugar, ANF, HB_gAg, anti-HCV, complement C₃; selective protein clearance.
- (C) Chest X-ray—May show evidence of hydrothorax or pneumonitis.
- (D) Ultrasonography of kidneys reveal normal, small or enlarged kidney size. Kidneys in glomerulonephritis are small while diabetic and amyloid kidneys are large in size.
- (E) Renal biopsy—For histological diagnosis, and also in assessing the treatment and prognosis.

How to diagnose that CRF has developed from nephrotic syndrome ?

1. Beginning of polyuria.
2. Diminution of anasarca.
3. Appearance of hypertension.
4. Development of anaemia.
5. Specific gravity of urine becomes low and fixed (1010).
6. Blood urea and creatinine levels increase.

Conditions associated with hypercholesterolaemia :

1. Pregnancy.
2. Diabetes mellitus (uncontrolled).
3. Myxoedema.
4. Nephrotic syndrome.
5. Obstructive jaundice.
6. Familial hypercholesterolaemia.
7. Prolonged intake of glucocorticoids, (3-blockers, thiazides and oral contraceptive pills).

Overall outcome of nephrotic syndrome :

The course may vary from complete recovery to death.

1. 30% recover completely.
2. 40% may die within a year or two after relapses and remissions (due to renal failure commonly).
3. 30% pass on to chronic renal failure.

* The long-term prognosis for minimal lesion nephropathy is excellent.

Common causes of CRF :

1. Chronic glomerulonephritis.
nephrotic syndrome.
2. Malignant hypertension.
3. Diabetes mellitus.
4. Chronic pyelonephritis.
5. Polycystic kidney disease.
6. Collagen vascular diseases
7. Obstructive uropathy.
8. Interstitial nephritis.
9. Analgesic nephropathy.
10. Renal tuberculosis.
11. Gout, multiple myeloma.

How do you like to manage your patient (minimal lesion) ?

1. To maintain an intake-output chart; intake = previous days urine output+ 500 ml.
2. High protein intake is not advised. Increased intake of protein may cause hyperfiltration in the living nephrons which may end in glomerulosclerosis and renal failure. Now-a-days, modest protein restriction (e.g., 0.6 mg/kg of body weight/day) is advocated, particularly in an azotaemic patient. Dietary proteins should be of high biological value and can be supplemented by amino acids.
3. No added salt in diet, i.e., 100 meq of sodium/day is allowed and often it may be as low as 30-50 meq/day.
4. Diuretics—Bendroflumazide 5-10 mg/day (in mild oedema), frusemide 80-120 mg/day or bumetanide (2-3 mg/day) or spironolactone (100-200 mg/day) in moderate oedema may be used singly or in combination (in severe oedema). The targeted weight loss should be <1kg/day as aggressive diuretic therapy may lead to ARF as a result of contraction in intravascular volume.
5. I.V salt-free albumin is given to increase the plasma oncotic pressure—20 g in 100 ml is used in 1 hour. It is costly and in hospitals, 'plasma' or blood transfusion is used for this purpose.
6. Corticosteroids and cytotoxic drugs—prednisolone 60 mg/m² in children and 1-1.5 mg/kg of body weight in adults is used daily for 4 weeks or until proteinuria disappears. After the urine is totally protein-free, the dose of prednisolone is gradually tapered to zero over next 4 weeks. But for those who show no response to initial 4 weeks' therapy, alternate-day prednisolone (35-40 mg/m² in children. 1 mg/kg in adults) for 4 additional weeks is advocated. Majority responds by the initial 4 weeks' treatment. Absence of response within 8 weeks is usually indicative of error in diagnosis and should be reviewed by a renal biopsy. Those who show relapse (within first year mainly), may be continued with low dose prednisolone i.e., 5-10 mg daily or on alternate days for 3-6 months. Those who have frequent relapses or show side-effects with prednisolone, may be given a brief course of cyclophosphamide (2-3 mg/kg daily) or chlorambucil (0.1- 0.2 mg/kg daily) daily for 2-3 months to induce subsequent remissions.
7. 'Statins' for the treatment of hypercholesterolaemia (e.g., atorvastatin 10 mg daily at bed time).
8. General measures—Multivitamins, antibiotics for secondary infection (as a result of hypogammaglobulinaemia), paracentesis of hydrothorax or ascitic fluid, metoclopramide for nausea and vomiting, packed red cells transfusion for anaemia, long-term oral anticoagulation in renal vein thrombosis, vitamin D supplementation in biochemical deficiency.
9. Emotional and psychological support to the child and his parents.
- [10. In other histological varieties—Steroids, cytotoxic drugs, anticoagulants and antiplatelet drugs are tried. Dialysis is advised if the patient goes into renal failure.]

Decoding of few terminologies in minimal lesion nephropathy :

1. Steroid resistant—Poor response with full dose of corticosteroid for 8 weeks.
2. Steroid dependent—When symptoms appear after withdrawal of steroid.
3. Steroid toxic—When side-effects of corticosteroid appear (like moon face, hypertension, peptic ulcer, stunted growth, behavioural changes etc). This is the commonly encountered complication while treating nephrotic syndrome.
4. Steroid responsive—Urine is free of protein within 3-4 weeks.

Conclusion :

1. Always measure the BP of the patient.
2. Take the H/O diabetes and hypertension routinely (past and family history).
3. Never forget to see the scrotal or vulval oedema, hydrocele, contact ulcers and phimosis. Examination of sacral oedema is mandatory.
4. Always percuss the urinary bladder for retention of urine (to make D/D with oligo-anuria).
5. Palpate the abdomen by 'dipping method' in the presence of ascites.
6. Examine for presence of hydrothorax (often bilateral).
7. Accurate drug history should be taken in all renal patients.

Section 6

LOCOMOTOR SYSTEM (OPTIONAL)

The locomotor system (or musculoskeletal system) comprises of muscles, bones, joints and soft tissue structures (e.g., ligaments and tendons).

Cardinal points in the history :

1. Onset, duration; pain and swelling of joint (arthritis) or only pain (arthralgia); axial or appendicular skeleton involved; big joints or small joints; mono-, pauci- or polyarticular; fleeting, additive or intermittent; morning stiffness, any pain during walk up and down the stairs, any associated H/O fatigue, weight loss, conjunctivitis, iritis, skin rash, skin nodule, mouth or penile ulcer, lymphadenopathy, alopecia, dry mouth, previous miscarriage.
2. H/O trauma, tuberculosis, bacillary dysentery, bleeding diathesis, enteric fever, sore throat, sexual exposure (STD-induced arthritis or reactive arthritis), fever, urethritis.
3. Family H/O tuberculosis, ankylosing spondylitis, haemophilia or gout; occupational history

Scheme of Examination

(A) Inspection :

- a) Mono-, pauci- or polyarticular — Joint/joints involved; 'attitude' of the limb.
- b) Swelling.
- c) Deformity (fixed or reducible; varus, valgus, ulnar, radial, flexion).
- d) Signs of Inflammation over the involved joint (e.g., erythema).
- e) Wasting of muscles.
- f) Skin changes.

(B) Palpation :

- a) Temperature of the local part (T warmth in inflammation, e.g., synovitis).
- b) Tenderness (in joint line or periarticular).
- c) Corroboration of the findings of inspection.
- d) Any swelling — Fluctuant or non-fluctuant; boggy feeling of thickened synovium.
- e) Muscle power e.g., grip strength.

(C) Movements (passive as well as active) :

- a) Restricted movement, instability or excessive mobility.
- b) Any pain on movement.
- c) Crepitus or grating sensation on movement.
- d) Any associated muscular spasm.

(D) Measurements ;

- a) Length and circumference of the limb.
- b) Measurement in relation to various body points.
- c) Thoracic ankylosis by chest expansion measurement and lumbar ankylosis by Schober's test.
- d) To measure joint movement with the help of goniometer.

(E) Examination of the spine for tenderness, mobility, kyphoscoliosis, lordosis, gibbus.

(F) Gait.

(G) Systemic examination—To search for evidences of pleurisy, pericarditis, vasculitis, hepatosplenomegaly, iritis, aortic incompetence or lymphadenopathy.

* Schober's test : It is performed to make out flexion movement of lumbar spine. The patient stands erect. Give two skin marks at the back, one 10 cm above and the other 5 cm below the lumbosacral junction (marked by a horizontal line joining the postero-superior iliac spines). On full forward flexion, these marks distract > 5 cm but no distraction of skin marks is seen in ankylosed spine.

** Read gout, different arthritis like rheumatic, rheumatoid, septic and osteoarthritis from standard text book. Vide the sections on 'Aortic stenosis', 'Examination of the hands' and 'Charcot joint' too.

Case 22

POLYARTHRITIS

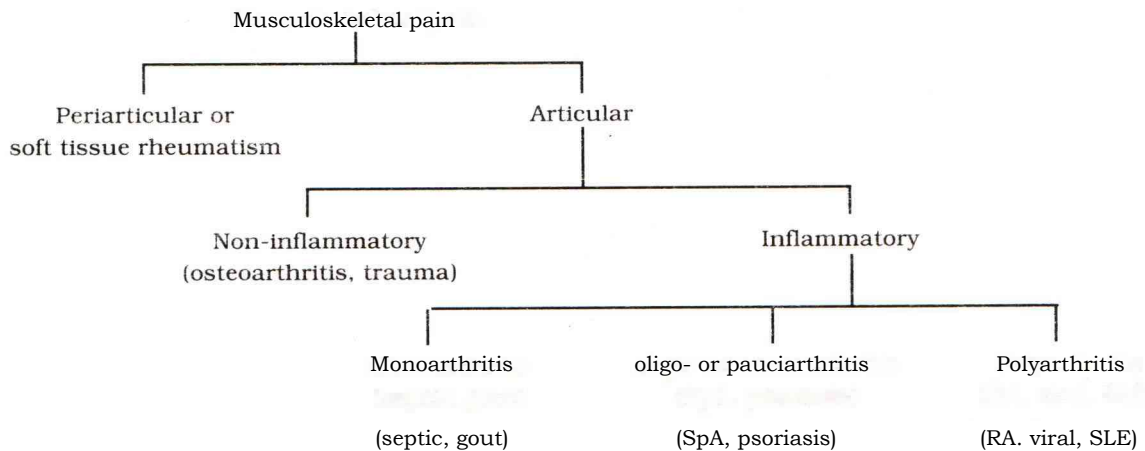
Clinical presentation :

A young female presents with H/O long-duration of joint pain, disability and deformity. She has bilaterally symmetrical involvement of small as well as large joints with active synovitis in few joints along with deformity of hands and feet. It is an additive arthritis of peripheral joints where the axial joints are not affected.

The most probable provisional diagnosis is rheumatoid arthritis.

How to approach in a patient of musculoskeletal (MSK) disorder?

Pain is the commonest manifestation and a near universal feature of MSK disorders. Since a wide variety of conditions give rise to MSK complains (i.e., arthralgia, myalgia, or bone pain), a systemic approach is necessary to make a provisional diagnosis. A careful history, meticulous clinical examination and judicious use of laboratory investigations help to arrive at a definitive diagnosis. The approach should be:



* RA - rheumatoid arthritis, SpA - spondyloarthropathy, ReA - reactive arthritis, SLE - systemic lupus erythematosus, CRP - C-reactive protein, ANA - antinuclear antibodies, RF - rheumatoid factor, MCP - metacarpophalangeal, MTP - metatarsophalangeal, PIP - proximal interphalangeal, DIP - distal interphalangeal, DMARDs - disease-modifying anti-rheumatic drugs, NSAIDs - non-steroidal anti-inflammatory drugs

Characteristics of inflammatory joint disorders :

Inflammatory joint disorders (RA, SpA, ReA) will have:

1. Significant early morning stiffness > 30 minutes.
2. Pain aggravation on resting the joints.
3. Symptomatic improvement on gentle use of joints.
4. Spontaneous flares (up-and-down course) are common.
5. Constitutional symptoms - fatigue, loss of appetite, weight loss, low-grade fever, night sweats are very often present.
6. Increased acute phase reactants e.g., high ESR and CRP.

* In non-inflammatory disorders, from 1 to 6, respectively they are <30 minutes, pain on moving the joints, no improvement, uncommon, absent, and normal.

What is soft tissue rheumatism (STR) ?

'Soft Tissue Rheumatism' is a term restricted to inflammatory or painful conditions which are, in general, non-articular in origin, and refers to aches or pains arising from structures surrounding the joint such as muscles, tendons, ligaments and bursae. This group accounts for the major chunk of musculo-skeletal disorders in any general outpatient clinic.

The anatomical basis of pain in MSK system could be due to:

Joint:

- Synovium - synovitis
- Joint capsule - capsulitis

Periarticular soft tissue:

- Bursa - bursitis
- Tendon - tendonitis
- Tendon sheath - tenosynovitis
- Insertion of tendon, ligament - enthesitis

Bone:

- Osteoporosis, osteomalacia, multiple myeloma, osteonecrosis

Differentiation of articular from non-articular/periarticular disorders ?

A careful clinical examination should be performed to differentiate soft tissue rheumatism from articular disorders. Bedside examination should follow:

1. Inspection: 'local' swelling around a joint (periarticular) favours STR rather than articular disorders whereas the joint (articular) swelling looks globular or fusiform surrounding a joint.
2. Palpation: In STR, tenderness is localised and usually away from joint-line margin. Articular disorders are conspicuous by the presence of joint-line tenderness, crepitus over the joint and/or joint effusion.
3. Movements: Active (the patient moves the joint) and passive (the physician moves the joint while the patient relaxes the limb) movements are equally impaired in articular disorders. In STR, pain is aggravated on active than on passive movement, and limitation of motion is always greater in active movement rather than passive one.

Common types of soft tissue rheumatism :

Shoulder: frozen shoulder, rotator cuff syndrome, bicipital tendonitis

Elbow (epicondylitis): tennis elbow, golfer's elbow

Bursitis: pre- and infrapatellar bursitis, olecranon bursitis, trochanteric bursitis

Tenosynovitis: De Quervains' tenosynovitis

Entrapment syndromes: carpal tunnel syndrome (median nerve), tarsal tunnel syndrome (posterior tibial nerve), meralgia paraesthetica (lateral femoral cutaneous nerve)

Pattern recognition in MSK disorders :

Pattern recognition in MSK disorders or rheumatological diseases is of immense importance to have a clean-cut diagnosis. Often features of two or more MSK disorders are present in a single patient to build up a diagnosis of 'overlap syndrome'. If the pattern of involvement is not recognized clinically, it is very difficult to clinch a diagnosis of rheumatological diseases only by ordering of RF, ANA, autoantibodies or other immunological markers.

1. Mode of onset: acute or insidious.
2. Duration of joint pain: acute (<6 weeks) or chronic (>6 weeks).
3. Number of joints affected: monoarthritis, oligo- or pauciartthritis, polyarthritis.
4. Pattern of involvement: axial (spine, sacroiliac, anterior chest wall, and shoulder and hip joint) or appendicular (peripheral joints). Shoulder and hip joints are known as root joints.
5. Distribution of joint involvement: symmetrical or asymmetrical, small or large joint, lower limbs or upper limbs, involvement of any specific joint (e.g., 1st metatarsophalangeal joint in gout, haemarthrosis of knee joint in haemophilia, DIP joints of hands in osteoarthritis).
6. Order or sequence of affection: intermittent (gout) or progressive (classical RA), migratory (rheumatic arthritis, SLE, serum sickness) or additive (RA, ReA).
7. Extra-articular manifestations: constitutional symptoms (e.g., fever), skin rash, subcutaneous nodule, oral ulcer, conjunctivitis or episcleritis, penile ulcer, nail changes (e.g., psoriasis), Raynaud's phenomenon (e.g., scleroderma).

Possible causes of polyarthritis, oligoarthritis and monoarthritis :

- (A) Polyarthritis - RA, viral arthritis (Parvovirus B19, hepatitis B, HIV, chikungunya), SLE, psoriasis, juvenile idiopathic arthritis (JIA), ReA, ankylosing spondylitis, systemic sclerosis, adult-onset Still's disease, generalized osteoarthritis, rheumatic arthritis, chronic gout, sarcoidosis (chronic), hypertrophic osteoarthropathy.
- (B) Oligo- or pauciartthritis - JIA, gout, SpA, psoriatic arthropathy, oligoarticular presentation of polyarthritis.
- (C) Monoarthritis - septic arthritis (S. aureus, N. gonorrhoea), crystal arthropathy (gout and pseudogout), tuberculous arthritis, osteoarthritis, SpA, traumatic, haemarthrosis (e.g., trauma, haemophilia), monoarticular flare of polyarticular disease, Charcot joint.

* **Few terminology** : Arthralgia - joint pain without obvious inflammation (i.e., only pain, no swelling), arthritis - pain and swelling in the joints, i.e., demonstrable features of inflammation; monoarthritis - affection of single joint, oligo- or pauciarthritis - affection of 2-4 joints, polyarthritis - involvement of 5 or more joints, synovitis - clinically apparent inflammation of synovium, enthesopathy / enthesitis - inflammation of an enthesis (i.e., the site of ligament, tendon, or articular capsule insertion into periosteum and bone), GALS screen - it is a quick reliable screen of gait, arms, legs and spine (GALS) of the locomotor system

Small and large joint involvement in MSK diseases :

- (A) Small joint arthropathy: RA, SLE, ReA, gout, nodular osteoarthritis, psoriasis, enteropathic arthritis (i.e., associated with inflammatory bowel disease or IBD, i.e., ulcerative colitis and Crohn's disease), sarcoidosis.
- (B) Large joint arthropathy: RA, ReA, ankylosing spondylitis, rheumatic arthritis, generalized osteoarthritis, psoriatic and enteropathic arthritis.
- (C) Both large and small joints arthropathy: RA, SpA, osteoarthritis.

Symmetrical versus asymmetrical involvement :

Symmetrical involvement refers to affection of same joints on both the sides though it may not be mirror-image symmetry. For example, involvement of left-sided 2nd and 3rd, and right-sided 4th MCP joints of hands in RA is deemed symmetrical involvement of MCP joints.

- (A) Symmetrical - RA, SLE, PSS, nodular osteoarthritis of hands, JIA and haemochromatosis
- (B) Asymmetrical - gout, SpA, metabolic arthropathy (acromegaly)

* Psoriasis may be symmetric or asymmetric at presentation.

Arthritis in relation to age and sex of the patient :

- (A) Age:
 - a) Children: JIA, haemophilia, trauma
 - b) Adolescents: RA, SpA, JIA, trauma, poststreptococcal reactive arthritis
 - c) Young: Trauma, gonococcal
 - d) Adults: SpA, ReA, SLE, psoriasis, gout
 - e) Middle age: RA, gout, osteoarthritis, scleroderma
- (B) Sex: Majority of arthritides are dominant in females except gout, ankylosing spondylitis, ReA (Reiter's syndrome) and polyarteritis nodosa which are common in males

Define rheumatoid arthritis and criteria for its diagnosis :

Rheumatoid arthritis is an autoimmune disorder of unknown aetiology characterized by chronic symmetrical polyarthritis, joint erosions and destruction. It is the commonest inflammatory joint disease seen in clinical practice, and in its world-wide distribution it affects 0.5-3% of the population. The course is extremely variable and sometimes associated with extra-articular features. RA is notorious for development of joint deformities, specially the small joints of hands and feet. The age of onset is commonly in between 30-50 years, and in gender predilection F:M = 3:1 before menopause while F:M = 1:1 post-menopause. RA is diagnosed by **ACR criteria** (American College of Rheumatology, 1987 revision), which are as follows:

- Morning stiffness > 1 hour
- Arthritis of 3 or more joints
- Arthritis of hand joints
- Symmetrical arthritis
- Rheumatoid nodules
- Rheumatoid factor
- Radiological changes (erosions and/or periarticular osteopenia)
- Duration of 6 weeks or more

* Presence of 4 or more criteria is necessary for diagnosis of RA

How a patient of rheumatoid arthritis presents ?

To diagnose a case as RA, one has to wait for 6 weeks (ref: ACR criteria). It starts with:

1. Aches and pains in the joints.
2. Insidious onset of polyarthritis.

3. Prolonged early morning stiffness.
4. Bilaterally symmetrical involvement.
5. Small joints of hands and (feet) are always involved in addition to large joint involvement. In small joints of hand (and feet), MCP (MTP) and PIP joints are commonly involved; DIP joints are virtually spared in RA. Large joints commonly involved are wrists, elbows, shoulders, knees and ankles.
6. Signs of inflammation may be seen in joints in the early part of the disease e.g., swelling (synovitis), erythema, warmth, and pain and tenderness.
7. As the disease progresses, deformity of hand and foot joints may develop in the absence of treatment.
8. The patient may suffer from different **extra-articular manifestations** along with joint involvement. They may be systemic (fever, weight loss, fatigue), musculoskeletal (tenosynovitis, bursitis, wasting of muscles), dermatological (nodules, pyoderma gangrenosum, leg ulcer, vasculitis), pulmonary (pleural effusion, bronchiolitis, fibrosing alveolitis, pulmonary nodules, Caplan's syndrome), cardiac (pericarditis, myocarditis, endocarditis, aortic regurgitation, conduction defects), ophthalmic (episcleritis, scleritis, keratoconjunctivitis sicca, scleromalacia), neurological (mononeuritis multiplex, cervical cord compression, entrapment neuropathy, peripheral neuropathy), lymphoreticular (anaemia, eosinophilia, thrombocytosis, Felty's syndrome, splenomegaly), and miscellaneous (amyloidosis, dry mouth, lymphadenopathy) involvement.

'A must' to remember in diagnosing rheumatoid arthritis :

A clinician should always remember that RA is:

- Entirely a clinical diagnosis.
- Typically bilaterally symmetrical.
- Classically presents as polyarthritis (may present as oligoarthritis).
- Don't diagnose if hands are not involved.
- DIP joint involvement is extremely rare.
- Lumbar spine is not involved in RA; cervical spine affection is very rare
- Presence of mucocutaneous rash, high pyrexia and active urinary sediments are against the diagnosis.

What is 'early' rheumatoid arthritis and what is its importance ?

It is the early disease having duration of persistent synovitis of around 3 months to 2 years. Different studies show that 10-26% patients of RA have radiographic bone erosion at 3 months, while 60% have it at 1 year, and 75% within 2 years. Unfortunately, bony erosions and deformities are largely irreversible. Hence, early disease represents a potential 'window of opportunity' for therapeutic interventions with DMARD, which has shown beneficial short and long term effects. So, present day goal is to diagnose RA as early as possible i.e., 'catch them young'. Early RA is diagnosed by RF, antibodies to cyclic citrullinated peptide (anti-CCP), high resolution ultrasonography (USG) with power and colour Doppler, and MRI scan of joints.

Describe different hand deformities in rheumatoid arthritis :

Read the section on 'Examination of hands'.

What are 'seronegative spondyloarthropathy' (SpA) ?

These are group of inflammatory joint diseases which are characterized by inflammatory low back pain in addition to peripheral arthritis. They suffer from asymmetrical oligoarthritis (lower limb affected more than upper limb), sacroiliitis and inflammatory spondylitis. There may have a positive family history and an **association with** HLA-B27 allele. The rheumatoid factor is always negative (seronegative). SpA encompasses 5 disorders like,

1. Ankylosing spondylitis (prototype of SpA),
2. Reactive arthritis (including Reiter's syndrome),
3. Psoriatic arthropathy,
4. Arthritis associated with inflammatory bowel disease (enteropathic arthritis), and
5. Undifferentiated SpA.

* The sixth group (according to some rheumatologists) is juvenile-onset SpA

Differentiate arthritis according to serology :

- (B) Seropositive (RF positive) - Sjogren's syndrome, RA, SLE, polymyositis-dermatomyositis, systemic sclerosis, sarcoidosis, vasculitis (polyarteritis nodosa). The higher the titre of RF, the more severe is the disease.
- (C) Seronegative (RF negative) - SpA (the whole group), gout, osteoarthritis, juvenile idiopathic arthritis (JIA; majority are RF negative), RA (30%), rheumatic arthritis, Still's disease:

* RF is also positive in certain non-rheumatic diseases like fibrosing alveolitis, leprosy, tuberculosis, infectious mononucleosis, syphilis, kala-azar, chronic hepatitis, infective endocarditis, cryoglobulinaemia, malignancy, and in normal population (5% women aged above 60 years).

Polyarthritis affecting hands — pattern of involvement :

1. Rheumatoid arthritis - MCP, PIP but spares DIP.
 2. Osteoarthritis - DIP but spares MCP; affects 1st carpometacarpal joint (base of thumb).
 3. Psoriatic arthropathy - commonly DIP.
 4. Chronic gout - MCP, interphalangeal joints.
 5. SLE - MCP joints commonly (Jaccoud's arthritis).
- DIP joints in hands are affected in osteoarthritis (Heberden's node), psoriasis, scleroderma, sarcoidosis, gout and adult-onset Still's disease.
 - PIP joints in hands are affected in RA, SLE, scleroderma and osteoarthritis (Bouchard's node).
 - MCP joints are commonly involved in RA, SLE, scleroderma and haemochromatosis.

Arthritis with nodule formation :

Rheumatoid arthritis (commonest), rheumatic fever and gout (tophi)

* Sarcoidosis (granuloma) and xanthoma (from hyperlipidaemia) may also have nodules.

What are Reiter's syndrome, Felty's syndrome, Still's disease and Caplan's syndrome ?

Reiter's syndrome — In the year 1916, Hans Reiter, the physician leader of the Nazi party in Germany during World War II, first described this syndrome. At present, the older term Reiter's syndrome is being used less frequently, and has been replaced by 'reactive arthritis'. The *classical triad of urethritis, conjunctivitis and arthritis* following a gastrointestinal (mainly Salmonella, Shigella, Campylobacter, Yersinia-induced) or genitourinary infection (non-gonococcal urethritis or sexually acquired infection with Chlamydia), previously referred to as Reiter's syndrome, is now considered part of the spectrum of reactive arthritis. The extra-articular manifestations are circinate balanitis (painless superficial ulceration of glans penis), keratoderma blennorrhagica (painless, red, raised pustules and plaques in sole of feet and palmar surface of hands), nail dystrophy (onycholysis and hyperkeratosis) and buccal erosions.

Felty's syndrome - It is a variety of long-standing rheumatoid arthritis which is common in females in the age group of 50-70 years, and is associated with splenomegaly, lymphadenopathy, neutropenia, and occasionally anaemia and thrombocytopenia. The patients remain strongly seropositive.

Still's disease - This is the seronegative variant of RA in children and rarely in young adults. The presenting features are high rise of swinging pyrexia associated with evanescent maculopapular rash, arthralgia and arthritis, myalgia and generalized lymphadenopathy. Hepatosplenomegaly and pleurisy / pericarditis are also evident. Anaemia, neutrophilia, thrombocytosis with high ESR and CRP are common. Serum ferritin may be enormously elevated in adult onset Still's disease.

Caplan's syndrome - This is the coexistence of seropositive rheumatoid arthritis and pneumoconiosis of the lung (shows rounded fibrotic nodules 0.5-5.0 cm in diameter in lung).

Common laboratory findings in rheumatoid arthritis :

No tests are specific for RA; however, following tests are commonly performed:

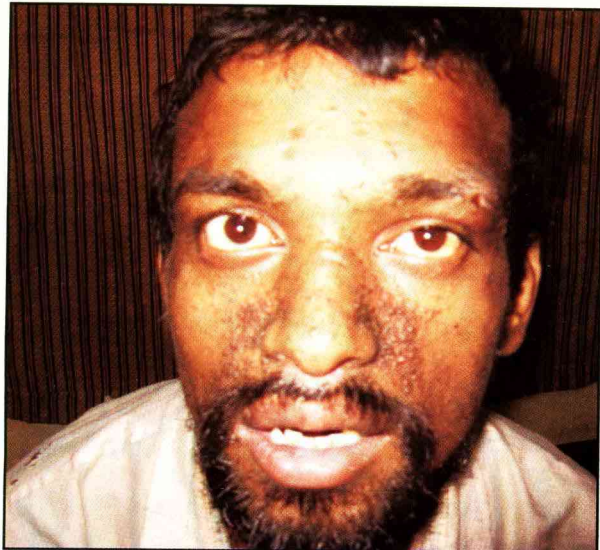
1. Anaemia (normocytic-normochromic), eosinophilia, thrombocytosis.
2. Raised ESR and CRP.
3. RF is positive in approximately 70% patients.
4. Positive antibodies to cyclic citrullinated peptide (anti-CCP) - having high specificity (90-98%); commonly done in seronegative patients to confirm the diagnosis of RA.
5. Acute phase response - High ferritin, reversed albumin/globulin ratio, moderate elevation of alkaline phosphatase. Raised ESR and CRP are also acute phase responses.
6. Radiology - Early RA may show only soft tissue swelling in X-ray. Stages of radiological (X-ray) progression in RA are periarticular osteopenia, marginal erosions, loss of articular cartilage.



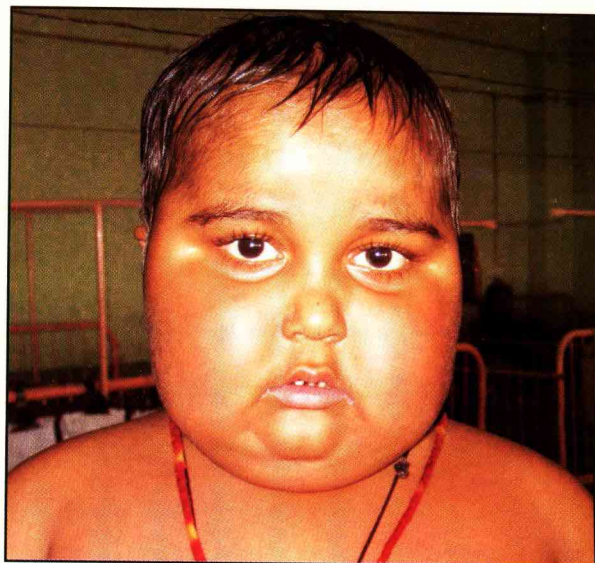
Short stature, webbed neck, low hair line, low set ear, 'shield-like' chest with mild mental subnormality in **Turner's syndrome** (girl aged 18 years)



Shadows of respiratory distress in face, central **cyanosis**, clubbing and **polycythemia** (suffused conjunctiva) in Fallot's tetralogy



Adenoma sebaceum, low intelligence and H/O epilepsy (injury in left eyebrow due to fall from convulsions) – classical triad in **tuberous sclerosis**



Moon face with facial plethora, excessive hair growth in forehead, double chin and obesity – an effect of long-continued steroid therapy



Jaundice, subconjunctival haemorrhage and dendritic ulcer in cornea – an AIDS patient with disseminated **herpes simplex virus** infection



Extensive **desquamation of palmar skin** during convalescence from toxic shock syndrome



Mycosis fungoides (cutaneous T-cell lymphoma) – multiple tumour like lesions in ulcerated base



Haemorrhagic blister, necrotising skin lesion, cellulitis and brawny induration – a complication of extravasation of cytotoxic drug while injected through intravenous canula

and subluxation and ankylosis. High resolution USG with colour and power Doppler picks up synovitis, effusion and erosion earlier than conventional X-ray. The MRI scan exhibit bone marrow oedema, synovitis and tendonitis. USG and MRI are rarely performed.

7. Joint fluid study (in effusion) - This is rarely performed. Due to presence of white cells, the aspirate often looks cloudy.

Hou> the disease activity is monitored in rheumatoid arthritis ?

This is assessed by,

1. Duration of early morning stiffness (measured by minutes).
2. Visual analogue scale for general health (eg, pain) as subjectively estimated by the patient.
3. Global assessment by observer as well as patient.
4. Joint count (number of swollen and tender joints).
5. Acute phase response - ESR and CRP.
6. NSAID pill count.
7. DAS (disease activity score) - a complex formula encompassing 28 tender joint count, 28 swollen joint count, ESR, and general health assessment by the patient on a visual analogue scale (VAS) from 0 to 100. It is measured by a DAS calculator.

Common imaging modalities done in musculoskeletal disorders :

1. X-ray: to demonstrate fracture, bony erosion, osteophytes, ankylosis etc.
2. Ultrasonography: to visualize soft tissue / periarticular structures / tendon. Baker's cyst, rotator cuff tear, thickened Achilles tendon.
3. Radionuclide scintigraphy: ^{99m}Tc -bisphosphonate (metastases, bone tumour, Paget's disease); ^{67}Ga -gallium and ^{111}In -indium scans are occasionally used to localize infections.
4. CT scan: for sacroiliitis, prolapsed intervertebral disc, trauma to the spine etc.
5. MRI scan: to visualize vessel, nerve, fascia, muscle, cartilage, ligament, synovial effusion, bone marrow oedema (e.g., early RA). Commonly used in spinal cord compression, prolapsed intervertebral disc, early avascular necrosis etc.
6. Bone mineral density (BMD) measurement: dual energy X-ray absorptiometry (DEXA) is the current method of choice to diagnose osteopenia and osteoporosis (T-score and Z-score).

Outline of management of rheumatoid arthritis :

The treatment paradigms of RA have shifted much from the past. In the past, traditional pyramidal approach (empirical) was go slow, go low but the present era is for early and universal use of DMARDs (evidence-based) though the future is waiting for combination therapy with DMARDs and biological agents (mechanism-based). Treatment needs coordination of a team of medical specialists (rheumatologist, orthopedic surgeon, physiatrist, physiotherapist, social worker, occupational therapist) who deliver an integrated programme of multidisciplinary care and rehabilitation. The **goal** is achieved by drugs, rest, physiotherapy, surgery, aids and appliances, and occupational and social services.

Aim of treatment - relief of symptoms, restoration of function and control of systemic involvement.

Different **modalities of management** are:

I. Pharmacological -

- a) NSAIDs (for relief of pain).
- b) Corticosteroids (for 'bridge therapy' before the onset of action of DMARDs, in rheumatoid flare, for extra-articular manifestations, and pregnancy).
- c) DMARDs - commonly used drugs are sulphasalazine, hydroxychloroquine, methotrexate and leflunomide. Less frequently used drugs are azathioprine, gold (oral and parenteral), cyclosporine A, chloroquine, cyclophosphamide, D-penicillamine, minocycline, chlorambucil and levamisole.
- d) Biological response modifiers (biologics) - these are genetically engineered treatment which basically acts by blocking TNF- α . The commonly used agents are etanercept, infliximab and adalimumab. Rituximab (causes lysis of CD-20 B cells) is also promising.

II. Surgical -

Decompression (in carpal tunnel syndrome), synovectomy, osteotomy, arthrodesis, arthroplasty, replacement of joints, joint fixation (atlanto-axial subluxation) etc.

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Section 7

LYMPHORETICULAR SYSTEM

The cardinal symptoms of lymphoreticular system are :

1. Painless or painful enlargement of lymph nodes
2. Pallor (resulting in weakness, palpitation, dizziness, ankle oedema, lassitude, anorexia, lightheadedness, breathlessness etc.)
3. Dragging pain or heaviness in the left hypochondrium (massive splenomegaly)
4. Haemorrhagic spots in the mucous membrane or skin
5. Epistaxis, gum bleeding, menorrhagia, haemarthrosis, H/O prolonged bleeding
6. Bone pain (multiple myeloma, acute leukaemia) and joint pain (Henoch-Schonlein purpura)
7. Jaundice (e.g., haemolysis)
8. Recurrent respiratory tract infection
9. Loss of body weight
10. Mass in the abdomen (abdominal lymphoma)
11. Sore throat (agranulocytosis), non-healing wounds (leucopenia)
12. Fever (leukaemia, lymphoma, neutropenia, infection with immunosuppression)

Scheme of Examination

1. Anaemia.
2. Haemorrhagic manifestations (haemorrhage into skin, conjunctival or buccal mucosa, epistaxis, gum bleeding, haemarthrosis, menorrhagia).
3. Oral cavity (gum for bleeding or hypertrophy, tongue, tonsil, involvement of Waldeyer's ring and skin (e.g., skin infiltration in lymphoma and leukaemia; telangiectasis).
4. Lymph nodes (in details).
5. Liver and spleen (in details).
6. *Sternal tenderness.*
7. Ophthalmoscopy or fundoscopy (for retinal haemorrhage, papilloedema, Roth spots etc.).

* This system is also known as **haemopoietic system** or **reticulo-endothelial system**.

N.B. : While examining haemopoietic system, examine the other systems for :

- I. CVS : haemic murmur and ejection systolic murmur for progressive anaemia, venous hum from severe anaemia, venous/arterial thrombosis from polycythemia vera.
- II. Nervous system : LMN type VIIth nerve palsy (leukaemic deposits), spinal cord compression from multiple myeloma, subacute combined degeneration from vitamin B₁₂ deficiency.
- III. Respiratory system ; Pleural effusion (e.g., lymphoma), pneumonia (due to immunocompromization).
- IV. Locomotor system : bone pain, joint swelling (haemophilia, gout).
- V. Genitourinary system : with special reference to examination of testes (ALL or NHL).
- VI. Skin and appendages ; pruritus (lymphoma), pigmentation (lymphoma, busulphan therapy in CML), leg ulcers (sickle cell anaemia, paraproteinaemia), splinter haemorrhage (acute leukaemia), koilonychia (iron deficiency anaemia).

Case 23

T HALASSAEMIA

What is your diagnosis ?

The diagnosis may be stated in one of the four ways. This is a case of,

1. Congenital haemolytic anaemia probably due to thalassaemia major, or
2. Moderate anaemia with hepatosplenomegaly probably from thalassaemia major, or

3. Huge splenomegaly probably due to thalassaemia major, or
4. Hepatosplenomegaly for discussion (if aetiological diagnosis can not be reached).

Why do you say so ?

This is a case of thalassaemia major because of the following :

- a) Young patient (18 years male patient) with positive family history, and there are presence of,
- b) Anaemia (moderate to severe).
- c) Jaundice (mild).
- d) Stunted growth.
- e) Mongoloid facies.
- f) Mild (to moderate) hepatomegaly with huge splenomegaly, and
- g) Past H/O repeated blood transfusions.

* **Features of chronic haemolytic anaemia are : anaemia, jaundice and splenomegaly.**

Chief complaints of your patient :

1. Respiratory distress since childhood and more for last 2 weeks.
2. Dragging pain in the left hypochondrium for last 4 years.
3. Yellowish discolouration of the eyes (from time to time) for last 5 years.
4. Repeated cough and cold, palpitation, ill-health and weakness since childhood.

What is the H/O present illness ?

1. Insidious in onset and gradually progressive illness.
2. Blood transfusion alleviates his sufferings.
3. Respiratory distress is exertional and often associated with purulent expectoration. There is neither any H/O PND nor any H/O seasonal variations.
4. Palpitation is exertional. There is no H/O fainting (black-out).
5. Dragging pain in the left hypochondrium is associated with a palpable mass in that region. Anorexia is profound.
6. Mild jaundice which increases from time to time.
7. There is no H/O gum bleeding, epistaxis, haematemesis, melaena or haemoptysis. The patient does not give any H/O leg ulcers or bipedal swelling. Occasionally he suffers from fever without any chill and rigor.
8. He has left school at 10 years of age. Till now he has been admitted several times in different city hospitals and each time he has been treated with blood transfusions by different doctors.

* Irregular fever in thalassaemia is due to increased metabolic activity and intercurrent infections.

What 'past history' you have enquired into ?

1. Whether the patient was admitted previously in any hospital for the same ailments.
2. Whether he has received blood transfusion during his past admissions in hospital.
3. Past H/O fever (chronic malaria and kala-azar).
4. Any H/O pain abdomen (pigment stone-induced or haemolytic crises).
5. Had he ever resided in Bihar or U.P of India (chronic kala-azar)?
6. As a routine the H/O diabetes mellitus, hypertension, rheumatic fever (joint pain, dyspnoea, palpitation), past history of jaundice, malaria or kala-azar (due to hepatosplenomegaly), tuberculosis should be enquired for.

* In thalassaemia, one should enquire the presence of similar disorder in brothers, sisters, parents and other family members. Is there any H/O death in the family due to jaundice?

What is a hereditary or familial disease ?

This is caused by ,

- a) Mutant genes.
- b) Follows the Mendelian law of inheritance and is transmitted from one generation to other.
- c) May not be present at birth.

Examples are—Diabetes mellitus, hypertension, bronchial asthma, schizophrenia.

What is a congenital disease ?

- a) Not produced by mutant genes.
- b) Does not follow Mendelian law of inheritance.
- c) Present since birth.

Examples are—Congenital heart disease, cleft lip.

Points noted in the treatment history of thalassaemia :

1. H/O blood transfusion.
2. Any specific drug used for the present disease (e.g., desferrioxamine).
3. Any surgery done in the past (pigment stone removal or splenectomy).

'General survey' in thalassaemia major :

In the 'general survey', the important points in favour of thalassaemia are :

1. Apparent age (often looks younger than actual age).
2. Build (there may be stunted growth).
3. Facies (mongoloid facies).
4. Anaemia (moderate to severe).
5. Jaundice (mild).
6. Pulse (may get water-hammer pulse due to severe anaemia).
7. Neck veins (may be engorged and pulsatile due to CCF),
8. Legs—Leg ulcer over malleoli (common in sickle cell anaemia),
9. Secondary sexual characters (there may be delayed development), and
10. Splenectomy scar (if there is absence of splenomegaly).

What is thalassaemia ?

This is an inherited (autosomal co-dominant) impairment of haemoglobin synthesis, in which there is partial or complete failure in the synthesis of a specific type of globin chain. It is a quantitative defect in the production of haemoglobin (qualitative defect is seen in sickle cell anaemia).

Thalassaemia is of two types :

- a) a (reduction or absence of α -chain synthesis), and
- b) β (reduction or absence of β -chain synthesis)—**Commonest amongst thalassaemic patients.**

(A) α -thalassaemia—4 types (seen in South East Asia, Middle East),

- (i) Silent carrier.
- (ii) α -thalassaemia trait.
- (iii) HbH disease.
- (iv) Hydrops fetalis (Hb Barts).

(B) β -thalassaemia—3 types (seen in mediterranean area),

- (i) Heterozygous (thalassaemia minor)—One of the parents has thalassaemia minor.
- (ii) Homozygous (thalassaemia major) or double heterozygous — Both parents have thalassaemia minor.
- (iii) Intermedia (homozygous who survive into adulthood are likely to have this milder form of disease. Haemolysis occurs but the patient is not transfusion-dependent).

* 'Intermedia' is more severe than 'minor' but milder than transfusion-dependent 'major'. Usually the patients suffer from moderate anaemia.

** HbE disorders and HbD Punjab are common in India.

Normal haemoglobin pattern in human being :**(A) Adult haemoglobin :**

- | | | |
|--------------------------------|---|--------------------|
| 1. HbA (97%) | — | $\alpha_2\beta_2$ |
| 2. HbA ₂ (1.5-3.5%) | — | $\alpha_2\beta_2$ |
| 3. HbF (< 2%) | — | $\alpha_2\gamma_2$ |

(B) Abnormal haemoglobin (in intrauterine life and neonates) :

- | | | |
|----------------|---|--------------------|
| 1 ■ HbF | — | $\alpha_2\gamma_2$ |
| 2. Hb Barts | — | γ_4 |
| 3. HbH | — | β_4 |
| 4. Hb portland | — | $Z_2\gamma_2$ |

* At term, HbF accounts for 70-90% of total Hb. HbF then falls rapidly and it becomes 25% at 1 month, and 5% at 6 month. The adult level (< 2%) may not be reached in some children until puberty.

** In an adult, the major haemoglobins seen are HbA and HbA₂ while in foetus, it is HbF and Hb Barts.

Why HbF is increased in thalassaemia major ?

Thalassaemia is an inherited abnormality in haemoglobin production, where there is partial or total

failure to synthesise a specific type of globin chain. In (3-thalassaemia major, there is reduction or absence of (3-chain production (defect within globin part) and thus, relative excess of α -chain synthesis occurs. Free α -chains have decreased solubility and will form inclusions within the RBC precursors in bone marrow. As a result of this, there is abnormality in membrane permeability as well as destruction of RBC by the macrophages. So thalassaemia major is characterised by both intramedullary erythroid destruction (ineffective erythropoiesis) and also a marked shortening of the life span of circulating RBC (peripheral haemolysis). Double injury to RBC results in expansion of red marrow (produces typical facial changes) and extramedullary haematopoiesis in the liver and spleen. There is also compensatory increase in synthesis of γ -chain which combines with excess of free α -chains and form a stable tetramer of HbF ($\alpha_2\gamma_2$). It is seen that patients with thalassaemia major who have relatively high rate of α -chain synthesis (i.e., more HbF), have less severe clinical course. A patient suffering from thalassaemia major may have HbF upto 98%. Thalassaemia major patients may prove fatal, if left untreated.

How do you classify haemolytic anaemia ?

(A) ACQUIRED—

- I. Environmental factors :
 - a) Antibody—Immune haemolytic anaemia (lymphoma, SLE, drugs).
 - b) Mechanical trauma—Microangiopathic haemolytic anaemia (prosthetic valves, haemolytic-uraemic syndrome).
 - c) Direct toxic effect—Malaria, clostridial infection.
 - d) Splenomegaly.
- II. Membrane abnormalities :
 - a) Spur cell anaemia (associated with portal cirrhosis).
 - b) Paroxysmal nocturnal haemoglobinuria (PNH).

(B) HEREDITARY:

- I. Membrane abnormalities :
 - a) Hereditary spherocytosis.
 - b) Hereditary elliptocytosis.
- II. Haemoglobinopathies :
 - a) Thalassaemia (quantitative defect).
 - b) Sickle cell disease (qualitative defect).
- III. Enzymopathies (metabolic defects) :
 - a) G₆PD deficiency.
 - b) Pyruvate kinase deficiency.

* Extracorporeal defects—all 'acquired' causes except PNH; Intracorporeal defects—all 'hereditary' causes plus PNH.

Describe thalassaemic or mongoloid facies :

in thalassaemia, there is \downarrow O_2 affinity of HbF resulting in tissue hypoxia \rightarrow which in turn, stimulates erythropoiesis \wedge expansion of marrow space gives rise to characteristic facial appearance.

1. Frontal bossing (prominent forehead due to marrow hyperplasia).
2. Depressed bridge of the nose (due to hyperplasia of lesser wing of sphenoid).
3. Hypertelorism (widely set eyes).
- 4 Apparent mongoloid slant of the eyes (mimics the eyes of Hindu Goddess Durga).
5. Malar prominence (due to marrow hyperplasia)—'Chipmunk' facies.
6. Dental malocclusion with prominent upper incisors (due to expansion of marrow).
7. Associated with mild icteric tinge of the conjunctiva, and pallor.

Why there is anaemia in thalassaemia major ?

1. Impaired haemoglobin production.
2. Ineffective erythropoiesis.
3. Haemolysis.
4. Haemodilution.
5. RBC destruction from hypersplenism.

Why there is jaundice in thalassaemia major ?

1. Haemolysis (classically it is haemolytic jaundice).

2. Viral hepatitis (type C or B infection due to repeated blood transfusions).
3. Rarely, due to iron deposition in liver.

Why there is cardiomegaly or heart failure seen in thalassaemia major ?

1. Persistent hypoxia (as progressive anaemia is always present).
2. Volume overload due to repeated blood transfusions.
3. Rarely, there is cardiomyopathy by haemosiderosis/haemochromatosis.

Description of splenomegaly in your patient :

1. Enlarged 16 cm below the left costal margin along its long axis (or along the splenic axis) and has crossed the umbilicus. It has enlarged towards the right iliac fossa.
2. Non-tender (rarely it may be tender due to splenic infarction, specially in a very big spleen).
3. A notch is present in the anterior border.
4. Firm in consistency.
5. It is freely moving with respiration.
6. Surface is smooth.
7. Margin is sharp.
8. Fingers cannot be insinuated between the spleen and the left costal arch.
9. It is neither bimanually palpable nor ballotable.
10. There is no colonic resonance over the mass.
11. No palpable splenic rub (present in case of splenic infarction).

* Never forget to auscultate over a hugely enlarged spleen for audible splenic rub.

Describe the hepatomegaly in your patient :

1. Enlarged 5 cm below the right costal margin at right MCL.
2. Moving with respiration.
3. Firm in feel.
4. Non-tender.
5. Smooth surface.
6. Margin is sharp and regular.
7. Left lobe is not palpable.
8. Upper border of liver dullness is at right 5th ICS at right MCL.
9. No pulsation, no rub, no bruit.

Splenohepatomegaly in thalassaemia is mainly due to extramedullary haematopoiesis and rarely transfusional haemosiderosis.

Features in other systems in thalassaemia major :

(A) CVS Effect of anaemia on CVS with special reference to haemic murmur. Read the section on 'Anaemia'. There may be features of heart failure. Look for neck veins carefully.

(B) RESPIRATORY SYSTEM—

1. Occasional rhonchi, and
2. Crepitations due to respiratory tract infection.

(C) NERVOUS SYSTEM— Rarely, there may be spastic paraplegia due to 'extramedullary haematopoiesis' in the paravertebral region in thorax.

(D) RETICULO-ENDOTHELIAL SYSTEM—

- a) Anaemia.
- b) Tonsils may be enlarged.
- c) No lymphadenopathy.
- d) No bleeding manifestations into skin.
- e) Hepatosplenomegaly.
- f> Rarely, sternal tenderness may be present (due to severe anaemia)

Ascites is not a feature of thalassaemia. In a **long case of thalassaemia**, first examine the G. I. system, next the CVS, respiratory system and so on. One may examine the reticulo-endothelial system after the examination of G.I. system.

How thalassaemia minor (trait) presents ?

They are rarely associated with significant clinical manifestations i.e., the patients can lead normal life with normal life expectancy. Usually they spend their life without diagnosis. 20% patients have splenomegaly and few may have pallor or mild icterus. The diagnosis is generally made when they are evaluated for persisting mild anaemia or pre-marital check up. Reticulocyte count is normal. HbA₂ is usually 5%; 50% patients have HbF elevated.

Differential diagnosis you will consider :

Basically one has to consider the **causes of huge splenomegaly**. For the discussion, read the section on 'Hepatosplenomegaly'. Actually, the task is to differentiate thalassaemia from chronic kala-azar, chronic malaria. CML. cirrhosis of liver (with hypersplenism) and tropical splenomegaly syndrome.

'Crises' described in haemolytic anaemias :

The 'crises' are very common in sickle cell anaemia. There are 4 types :

1. Haemolytic crisis (sudden hepatosplenomegaly with rapidly developing anaemia).
2. Aplastic crisis or aregenerative crisis (temporary marrow suppression)—Occurs in the presence of infection and/or folic acid deficiency, and may be seen even in thalassaemia major.
3. Vaso-occlusive crisis (commonest)—Acute and severe bone pain results from plugging of small vessels in bone. Various part of the body like hands and feet in children (resulting in dactylitis), or humerus, femur, ribs, vertebrae and pelvis in adults may be affected. Sweating, pyrexia and tachycardia are associated with.
4. Sequestration or infarction crisis—Thrombosis of the venous outflow results in crisis in an organ. Splenic infarction in sickle cell anaemia is a classical example.

* In haemolytic crisis, reticulocyte count increases but in other crises reticulocyte count does not increase from the basal level. By about 8 years of age, in sickle cell disease, spleen is no longer palpable due to repeated episodes of splenic infarcts leading to 'autosplenectomy'.

Pain abdomen in thalassaemia major :

1. Dragging pain due to huge splenomegaly.
2. Vaso-occlusive crisis (rare).
3. Biliary colic (due to production of pigment stones from haemolysis).
4. Rarely due to splenic infarction.

How to establish the diagnosis of thalassaemia major ?

1. Peripheral blood smear—
 - (i) Anisocytosis (variation in size), poikilocytosis (variation in shape), **microcytic hypochromic anaemia, target cells**, tear-drop cells, cigar-shaped cells, basophilic stippling, polychromasia, normoblasts. Small number of immature myelocyte or metamyelocyte may be present.
 - (ii) **Reticulocytosis** (indicates haemolysis and increased production of new red cells; normal reticulocyte count is 0.2-2% of RBC).
2. Haemoglobin concentration, ESR and bilirubin —
 - (i) Hb—Low (usually between 2-6 g/dl).
 - (ii) ESR—High.
 - (iii) Serum bilirubin is raised (unconjugated hyperbilirubinaemia).
3. Radiology—
 - (i) **X-ray of skull—'Hair on end' appearance** due to the separation of two tables of skull (i.e., widened diploic space) with perpendicular trabeculae in between.
 - (ii) **X-ray of small bones of hands—'Mosaic-patterns'** due to widening of medullary space with thinning of cortex and criss-cross trabeculae in between.
 - (iii) Chest X-ray (PA view) —Cardiomegaly, pneumonitis or paravertebral shadow as a result of extramedullary haematopoiesis.
4. **Haemoglobin electrophoresis is diagnostic**—Shows presence of **HbF (> 2% of total Hb)** and variable amount of HbA₂ and HbA (HbA₂ is increased more in thalassaemia minor than in major, and is usually > 3.5%).
5. Alkali denaturation test — HbF is resistant to alkali denaturation (i.e., the test is positive).
6. Osmotic fragility test—It is decreased, i.e., there is increased resistance of red cells to osmotic lysis (osmotic fragility is increased in hereditary spherocytosis).
7. Ferrokinesis—**Serum iron concentration is increased**, and TIBC usually remains normal or reduced.

8. Red cell survival study.
9. Bone marrow—Hyperplastic bone marrow with hyperplasia of erythropoietic and leucopoietic tissue. Myeloid and erythroid ratio (M : E) becomes 1 or less. Granulopoiesis and thrombopoiesis are preserved.
10. Both the parents have thalassaemia minor.

N.B. : The other names of thalassaemia major are Cooley's anaemia, target cell anaemia, mediterranean anaemia (*Greek 'thalassa' means sea*) and splenic anaemia.

Laboratory features of haemolysis :

1. **High serum bilirubin, usually < 6 mg/dl (unconjugated > conjugated).**
2. **High urinary urobilinogen.**
3. Low plasma haptoglobin and haemopexin.
4. Haemoglobinaemia and haemoglobinuria.
5. Increased methaemalbumin in blood (detected by Schumm test).
6. High urinary haemosiderin.
7. Fragmented red cells in peripheral blood.
8. **Reticulocytosis.**
9. Hyperplastic bone marrow (i.e., erythroid hyperplasia).

* 4,5 and 6 are classically seen in intravascular haemolysis.

Features of iron overload in thalassaemia major :

Here, iron overload is due to :

- a) Haemolysis, and
- b) Repeated blood transfusions.

Features of iron deposition (haemosiderosis) in different systems are :

1. CVS—Pericarditis, cardiomyopathy, ventricular ectopics, heart blocks, pump failure.
2. Hepatic—Hepatomegaly (prothrombin time and liver enzymes may increase), rarely cirrhosis of liver.
3. Endocrine—
 - (i) Pancreas—Diabetes mellitus.
 - (ii) Adrenal gland—May have adrenal crisis.
 - (iii) Due to diminished gonadotropin release— Delayed development of secondary sexual characters (due to impaired hypothalamo-pituitary-gonadal function), i.e., features of hypogonadism.

* Liver biopsy in thalassaemia major shows both reticulo-endothelial and parenchymal iron.

Causes of iron overload in internal medicine :

1. P-thalassaemia major
2. Haemochromatosis
3. Sideroblastic anaemia
4. Alcoholic liver disease
5. Repeated blood transfusions
6. Iron-dextran infusions
7. Chronic haemodialysis.

Complications of thalassaemia major :

1. Stunted growth with facial disfigurement.
2. Pigment gall stones.
3. Iron overload complications (described above)—Haemosiderosis.
4. Leg ulcers.
5. Hypersplenism.
8. Repeated respiratory tract infection.
7. Compressive myelopathy (due to extramedullary haematopoiesis).

Table 16 ; Differences between haemosiderosis and haemochromatosis

Features	Haemosiderosis	Haemochromatosis
1. Inheritance	1. Always acquired	1. Genetic or acquired
2. Tissues affected	2. R-E cells	2. Parenchymal cells
3. Damage	3. Less tissue damage	3. Obvious tissue damage
4. After desferrioxamine application	4. Urinary excretion of iron does not exceed 4 mg/24 hrs	4. Exceeds 4 mg/24 hrs

Causes of microcytic hypochromic anaemia :

1. Iron deficiency anaemia.
2. Thalassaemia.
3. Sideroblastic anaemia (often responses to pyridoxine).
4. Sometimes seen in lead poisoning.
5. Anaemia of chronic diseases (e.g., rheumatoid arthritis).

* Sideroblast—These are nucleated RBC having excess of iron-containing granules in the cytoplasm.

MCV, MCH, MCHC in thalassaemia :

1. Normally MCV is 87 ± 7 cubic (low in thalassaemia).
2. Normally MCH is 29.5 ± 2.5 pico-gram (low in thalassaemia).
3. Normally MCHC is 35 ± 3 g/dl (low in thalassaemia).

Severe anaemia without splenomegaly—possibilities in clinical medicine :

1. Anaemia due to acute or chronic blood loss.
2. Aplastic anaemia.
3. Sick cell anaemia (spleen shrinks due to repeated infarction).
4. Anaemia due to hookworm infestation.
5. Chronic renal failure.

What is the treatment of thalassaemia major ?

1. **No oral or parenteral iron therapy.**
2. **Repeated transfusions** to keep the haemoglobin in the range of 10 g/dl with desferrioxamine therapy.
3. Folic acid—5 mg daily, orally.
4. Antibiotics for infections.
5. Splenectomy in some cases.

* Thalassaemia minor needs no treatment. The patient should be reassured and genetic counselling is explained to the patient.

Recent trends in the management of thalassaemia major :

To maintain the most beneficial Hb level, it requires an individualised programme of transfusion with iron chelation and careful attention to the development of skeletal, endocrine and cardiac abnormalities.

1. Transfusion therapy—
 - (i) **Hypertransfusion**—Hb level is kept > 10 g/dl.
 - (ii) **Supertransfusion**—Hb level is kept > 12 g/dl.
2. Iron chelation is achieved by desferrioxamine which delivers drug by a small portable pump. The dose is 20-40 mg/kg/day and is given by subcutaneous continuous infusion. 1 g of desferrioxamine chelates 85 mg of iron. Now-a-days, It is started at 5 years of age when the child can use the 'infusion pump' of his own. Infusion lasts 10-12 hours (overnight) and is given for 5-7 nights per week with a goal to keep serum ferritin < 1000 ng/ml. Common side effects are pruritus, local swelling, sore arm etc. Excessive doses may give rise to cataract, optic neuritis, nerve deafness and retinal damage.
3. Splenectomy (see below) which is preceded 4-6 weeks by pneumococcal, H. influenzae and meningococcal vaccine.
4. Vitamin E—Can be used as an antioxidant drug; excretion of iron is enhanced by vitamin C (low dose, i.e., usually 3 mg/kg daily is used to avoid carcinotoxicity).
5. Vitamin D and calcium may be used to control osteoporosis.
6. Local irradiation of face to minimise facial disfigurement.
7. Neocyte transfusion i.e., transfusion of young low-density RBC (average life span 90 days); the neocytes reduce the rate of iron accumulation. Gerocytes (old RBC) may be removed by pneresis to reduce iron load.
8. Genetic manipulation is done by azacitidine or butyrate which Increases the production of HbF (increased HbF in thalassaemia Is associated with less severe clinical course).
9. Allogenic bone marrow transplantation from HLA compatible sibling (definitive therapy).
10. Folic acid supplement and antibiotics, as and when necessary. Chronic leg ulcers may be managed with elevation of the affected leg and daily dressings with zinc sulphate.
11. Prevention is possible by Identifying a foetus with p-thalassaemia by DNA analysis of chorionic villus in early pregnancy (8-10 week) and also by amniocentesis (14-18 week). Fetal cord blood

In the womb can also be tested In 18-20 weeks of pregnancy. This is appropriate if both the parents are known to be carriers, i.e., β -thalassaemia minor, and always terminate the pregnancy in these cases. Genetic counselling is a must.

- * In both the parents are thalassaemia minor :
 - 25% chance to have thalassaemia major
 - 50% chance to have thalassaemia minor
 - 25% chance to have a normal child

Advantages of hyper- and supertransfusion :

1. Gastrointestinal iron absorption is less.
2. Growth and development are increased.
3. Less chance of facial disfigurement.
4. Less chance of hypersplenism.
5. Less need for early splenectomy.

Indications for splenectomy in thalassaemia major :

1. Mechanical problems due to massive splenomegaly (dragging pain, early satiety).
2. Requirement of excessive transfusions (normal requirement is 1 unit / 4-6 weeks to maintain the Hb level in the range of 10 g/dl) e.g., double of normal requirement.
3. Features of hypersplenism.

* Splenectomy should be done in a child who is > 5 years of age.

** If spleen is not palpable in thalassaemia, always look for the splenectomy scar.

Peripheral blood film after splenectomy :

1. Marked anisocytosis, poikilocytosis.
 2. Howell-Jolly bodies.
 3. Target cells.
 4. Thrombocytosis.
 5. Pappenheimer bodies (sideroblastic granules).
 6. Heinz bodies.
 7. Nucleated RBCs.
 8. Basophilic stippling.
- * Remnants of nuclear material in RBC; ** Formed from denatured aggregated Hb.

Indications (medical) of repeated blood transfusions in your hospital :

1. Thalassaemia.
2. Aplastic or hypoplastic anaemia.
3. Haemophilia.
4. ITP (immune thrombocytopenic purpura).
5. Recurrent haematemesis or melaena.
6. Chronic renal failure.

Drawbacks of repeated blood transfusions :

1. Risk of iron overload and heart failure.
2. Chance of isoimmunisation.
3. Entry of infections through blood like malaria, hepatitis C and B; AIDS, toxoplasmosis, brucellosis, cytomegalovirus infection, syphilis; citrate toxicity.

* Acute (< 72 hrs) complications are anaphylaxis, transfusion reactions (chills, urticaria, breathlessness etc), febrile reactions, fluid overload resulting in LVF, mismatch (haemolysis), hypothermia. Delayed (> 72 hrs) complications are thrombophlebitis and entry of different infections.

Causes of target cell in peripheral blood film :

1. Thalassaemia.
2. Iron deficiency anaemia.
3. Cholestatic jaundice.
4. After splenectomy.

Common causes of refractory anaemia in clinical practice :

1. Aplastic anaemia.
2. Sideroblastic anaemia (pyridoxine-responsive).
3. Erythroleukaemia (M_e variety of AML).
4. Aleukaemic leukaemia.
5. Myelodysplastic syndrome.
6. Chronic renal failure.

Conclusion :

H/O repeated blood transfusions and previous hospital admissions should always be enquired. Menstrual history should be taken with care as in many patients menarche may not be attained. Never forget to examine for cardiomegaly, CCF and systolic murmur over the precordium.

Case 24

CHRONIC MYELOID LEUKAEMIA

What is your diagnosis ?

The diagnosis may be delivered in one of the three ways :

1. This is a patient of huge splenomegaly and moderate hepatomegaly probably due to chronic myeloid leukaemia (CML), or
2. This is a patient of anaemia with hepatosplenomegaly probably due to chronic myeloid leukaemia, or
3. Huge splenomegaly probably due to chronic myeloid leukaemia.

Why do you say so?

This is a case of CML because of the following :

1. This is an adult patient (majority are in the age group of 30-80 years with peak age at 55 years) who complains of gradual onset of dragging pain in the abdomen, mainly the left upper quadrant with a palpable, non-tender swelling present there. This is associated with loss of appetite and early satiety, vague ill-health, abdominal fullness, tiredness, marked weight loss and occasional rise of temperature with night sweats but without chill and rigor for last 5 months. The onset of the disease is insidious and the progress is slow.
2. There is neither any past H/O haematemesis, melaena, jaundice, blood transfusion, oedema feet nor it is associated with positive family history with similar type of illness.
3. On examination there is mild anaemia, absence of jaundice, moderate hepatomegaly with huge splenomegaly as well as there is presence of sternal tenderness. Lymphadenopathy, signs of hepato-cellular failure or bleeding manifestations into skin are absent.

Chief complaints of your patient :

1. Gradually progressive swelling with dragging pain in the left upper abdomen, specially in up-right posture for last 5 months.
2. Loss of appetite, tiredness, weakness and loss of weight for the same duration.

Presence of sternal tenderness—conditions associated :

Clinical method—Press the upper part of the body of sternum with the **ball of the right thumb** for 2-3 seconds. In the presence of sternal tenderness, the patient winces with pain (also see the facial grimace) or complains of pain. Sternal tenderness is found in :

1. Acute leukaemias (AML and ALL).
2. Chronic myeloid leukaemia.
3. Severe anaemia due to any cause (except aplastic anaemia).
4. Multiple myeloma.
5. Rarely in osteomalacia and osteoporosis.
6. After sternal puncture.

N.B. : When sternal tenderness is present, examine the patient for tenderness in other bones like pelvic bones, ribs, vertebrae, long bones (press the upper part of shin bone) and frontal bone (press the forehead). Look for the evidence of sternal puncture (benzene stain or cotton seal in the sternum) in these cases. Sternal tenderness is usually due to *proliferation as well as hypercellularity of bone marrow*.

Remember, diffuse bone pain is common in osteomalacia and multiple myeloma.

* 1 and 2 are most common causes.

Causes of swelling in the left upper abdomen :

- | | |
|-------------------------------|--|
| 1. Splenomegaly. | 5. Tumour of left adrenal gland. |
| 2. Kidney lump (left). | 6. Rarely, tumour of the tail of pancreas. |
| 3. Tumour of splenic flexure. | 7. Enlarged left lobe of liver. |
| 4. Growth from stomach. | 8. Omental mass. |

'Past history' you have enquired for :

- | | |
|--|--|
| 1. Haematemesis or melaena. | 6. Ankle oedema. |
| 2. Jaundice. | 7. Bleeding manifestations, e.g., purpura. |
| 3. Fever (ref : malaria or kala-azar). | 8. Haematuria (ref : kidney lump). |
| 4. Repeated blood transfusions. | 9. Diabetes mellitus. |
| 5. Bone pain. | 10. Systemic hypertension. |

Causes of huge splenomegaly :

Read the section on 'Hepatosplenomegaly'.

What is the classical description of the spleen of CML :

Splenomegaly in CML is present in 90% cases and approximately in 10%, the enlargement is massive and the spleen goes towards the right inguinal ligament. Splenic rub may be present.

For typical description—Read the section on 'Thalassaemia' as the description of splenic enlargement is the same in both the conditions.

* The spleen of CML is very hard and is known as '*cast iron spleen*'. Hepatomegaly occurs in 50% cases.

Phases of CML :

1. Chronic or stable phase—Easily controlled by treatment. Usually lasts for 3-5 years.
2. Accelerated phase—Control of the disease is difficult (progressive anaemia, organomegaly, fever, thrombocytosis or thrombocytopenia develop).
3. Blastic (blast crisis) phase—It is a dreadful condition where the patient transforms into acute leukaemia (AML in 70% and ALL in 30%). **The blast cell of AML contains Auer rods which are not seen in blast crisis of CML.** This is the cause of death in majority of patients.

* CML is a monoclonal stem cell disorder or myeloproliferative disorder characterised by an overproduction of myeloid cells with fairly normal maturation.

Why it is not a case of chronic malaria, chronic kala-azar or thalassaemia ?

Read the sections on 'Hepatosplenomegaly' and 'Thalassaemia'.

Why it is not a case of chronic lymphocytic leukaemia (CLL) ?

The salient features of CLL are :

1. Peak age for the disease is 65 years.
2. Onset is very insidious.
3. Anaemia develops slowly and lately.
4. Absence of sternal tenderness.
5. Presenting complaint is painless, discrete, firm and moderate enlargement of lymph nodes all over the body.
8. Moderate splenomegaly.

These features are absent here and this is why it is not a case of CLL.

How will you recognise the 'blast crisis' ?

The features of blast crisis are :

1. The stable patient will deteriorate (there is refractoriness to treatment).
2. Anaemia progresses rapidly; bone pain and sternal tenderness increase.
3. Severe bleeding starts e.g. petechiae, purpura, bruise, epistaxis, gastro-intestinal bleeding, bleeding from urogenital tract; fever appears.
4. Generalised lymphadenopathy may be present (due to transformation into ALL).
5. Abrupt increase in splenic size and tenderness.
6. Peripheral blood film shows > 10% blast cells.

Manifestations at the late stage of CML :

1. Bleeding into the skin or from mucous membrane of internal organs.
2. Acute pain in the left hypochondrium—Due to splenic infarction with audible splenic rub.
3. Persistent rise of temperature.
4. Leukaemic infiltrations in cranial nerves with their paralysis, pleural effusion, ascites or peripheral neuropathy.
5. Respiratory tract infection.

Differential diagnosis you will consider in this case :

Consider causes of massive splenomegaly.

How your diagnosis will be established ?

1. Blood examination—
 - a) Hb and RBC—RBC count is low with anisocytosis, poikilocytosis, few normoblasts, normocytic normochromic anaemia, low Hb (mean Hb level is 10.5 g/dl).

b) WBC—

(1) Average total count (TC) Is 1.5 lacs to 2.5 lacs /mm³ (1 to 5 lacs/mm³).

(ii) Differential count (DC) includes myelocytes 20-30%, metamyelocytes 25%, promyelocytes 2-3%, polymorphs 40%, eosinophils 4%, basophils 2%, lymphocytes 5%, monocytes 1%, myeloblasts 1-2% (usually < 10%).

c) Platelets—Initially normal or increased but diminished gradually.

N.B. : In the accelerated phase, more primitive cells appear and in the blastic phase, there is dramatic increase in blast cells (>10%) in the peripheral blood film.

d) ESR—Increased.

2. Bone marrow examination—It is not mandatory (though gives definitive diagnosis) because **peripheral blood picture is the most useful diagnostic aid**. It shows increased myelocyte series with increased myeloid and erythroid ratio (as high as 20:1). Often prominent megakaryocytes are seen (In acute leukaemias, bone marrow is hypercellular and there is presence of more than 30% of leukaemic blast cells). Cytogenetics show t(9;22) translocation.
3. Philadelphia (ph) chromosome is seen in myeloid and erythroid series of blood and bone marrow.
4. Other findings-
 - a) High serum uric acid and LDH level (due to increased cell breakdown).
 - b) High serum vitamin B₁₂ level (due to T in transcobalamin III present in neutrophil granules).
 - c) Low leucocyte alkaline phosphatase (LAP score).
5. Special investigation—Fluorescein-in-situ hybridization (FISH) or reverse transcriptase polymerase chain reaction (RT-PCR) helps demonstrating cytogenetic/molecular abnormality.

* A hypermetabolic state is found in CML, lymphoma, tuberculosis, malignancy and myelofibrosis.

** LAP score is diminished in CML and paroxysmal nocturnal haemoglobinuria. It is increased in infection (leukaemoid reaction), pregnancy, after steroid administration, stress, Hodgkin's disease, polycythemia vera and myelosclerosis.

*** The mean haemoglobin level is 10.5 g/dl (i.e., mild anaemia). Anaemia in CML is never severe in degree. Basophilia often points towards the diagnosis and tends to increase as disease progresses.

**** RNA analysis in CML demonstrates the presence of the chimeric BCR-ABL oncogene. ABL stands for abelson (named after the abelson murine leukaemia virus), and BCR for breakpoint cluster region (BCR gene on chromosome 22).

Hypercatabolic (hypermetabolic) symptoms of CML :

- | | |
|---------------------------|--------------------------|
| 1. Weight loss (wasting). | 4. Malaise and anorexia. |
| 2. Fever. | 5. Gout. |
| 3. Sweating (night). | 6. Heat intolerance. |

What is philadelphia (Ph) chromosome ?

1. Diagnostic of CML.
2. Shortening of long arms of chromosome G 22 (due to balanced reciprocal translocation of genetic material with chromosome 9).
3. Found in 90% cases of CML. Thus, 10% patients are Ph-chromosome -ve.
4. Ph-chromosome +ve cases have a better prognosis than Ph-chromosome -ve cases.
5. Found in myeloid and erythroid series of blood and bone marrow (never seen in lymphocytes).
6. Present in all the phases throughout the course of the disease and even after therapy; however Ph +ve cells decrease with a-interferon and imatinib therapy.
7. Helps in differentiation between AML and blast crisis of CML.

* By polymerase chain reaction (PCR), Ph-chromosome is present in almost all patients of CML.

Confirmation of diagnosis of CML :

1. Peripheral blood film with bone marrow examination.
2. Ph-chromosome—Pin-points the diagnosis.
3. RNA analysis for presence of BCR-ABL oncogene.
4. Low LAP score.

Aleukaemic and subleukaemic leukaemia :

In acute leukaemia, an increase in total WBC count is expected but in —

- a) Aleukaemic leukaemia—The total WBC count remains normal or subnormal, and there is absence of abnormal cells in the peripheral blood.

- b) Subleukaemic leukaemia—The total WBC count is within or below normal limit but few immature cells are seen in the peripheral blood.

Both a) and b) present as acute leukaemia. The confused **diagnosis is confirmed by bone marrow examination.**

What is juvenile CML ?

This rare form of CML is characterised by :

1. Affection below 4 years of age.
2. Absence of Ph-chromosome.
3. Increased level of HbF.
4. Normal cytogenetic karyotype.
5. Paucity of blastic transformation.
6. Presence of prominent monocytosis and thrombocytopenia.

What is myelodysplastic syndrome (MDS) ?

These are a group of acquired clonal abnormalities of haematopoietic stem cells with varying degrees of cytopenia which represent steps in the pathway of development of leukaemia. Often it is called 'preleukaemia' or oligoblastic leukaemia as these patients are at increased risk of developing acute leukaemia. People over 50 years of age are usually affected (median age 69 years). Blast cells in marrow are increased but never cross 30% (> 30% blast cells in marrow indicates acute leukaemia). Bone marrow examination shows hypercellularity with evidence of dysplasia. Though the exact cause is unknown, exposure to benzene or chemotherapeutic agents, or radiation may lead to MDS. Chromosomal abnormality is seen in up to 80% of cases. There is high morbidity and mortality due to bone marrow failure (symptoms of anaemia, bleeding or recurrent infections) or transformation into acute leukaemia.

The *Five distinct clinical syndromes* under MDS (FAB subtypes) are refractory anaemia, refractory anaemia with excess of blasts (RAEB), refractory anaemia with ring sideroblasts (RARS), chronic myelomonocytic leukaemia (CMML), and refractory anaemia with excess of blasts in transformation (RAEB-t). The WHO classification is a bit different. In the combined (FAB and WHO) classification, CMML is not included in MDS.

Prognosis of CML :

1. Median survival of treated CML is 3 years (in imatinib era, it is 4 years).
2. Median survival is about 1 year after blast crisis and < 1 year in Ph-ve variety.
3. Few patients survive over 10 years.

Complications of CML :

1. Blast crisis.
2. Haemorrhage into skin or mucous membrane of internal organs.
3. Recurrent infection (commonly in respiratory tract).
4. Hyperuricaemia (C/O joint pain due to gout).
5. Leukaemic infiltrations in cranial nerves, pleura (effusion), bones (paraplegia).
6. Rupture of the spleen.
7. Infarction of the spleen.
8. Priapism (persistent painful erection of penis).

* 'Leucostasis' in CML may produce priapism, blurred vision or respiratory distress.

Physical findings in different systems in CML :

- (A) GENERAL SURVEY—Mild anaemia, emaciation; pedal oedema due to anaemia or pressure over IVC by big spleen, haemorrhagic spots into skin may be present; very rarely jaundice may appear due to hepatic involvement, or type B or C hepatitis from blood transfusion.
- (B) G.I. TRACT—Huge splenomegaly, mild to moderate hepatomegaly. Usually there is absence of ascites. *The hallmark physical finding in CML is huge or massive splenomegaly.*
- (C) LYMPHORETICULAR SYSTEM—Presence of sternal tenderness; rarely lymphadenopathy in blastic transformation into ALL. Bone pain due to infiltration of marrow by leukaemic cells may be present.
- (D) RESPIRATORY SYSTEM—Basal crepitations due to pulmonary congestion; pleural effusion or rarely pneumonic consolidation.

- (E) CVS—Rarely, pericardial effusion.
- (F) NERVOUS SYSTEM—Cranial nerve involvement (search for Bell's palsy), paraplegia; rarely meningeal signs may be seen. Retinal haemorrhage due to leucostasis may be evident on fundoscopy.
- (G) GENITOURINARY SYSTEM—Sometimes there is presence of priapism.

Table 17 ; Bedside differentiation between acute leukaemias

Features	AML	ALL
1. Symptoms	Fever, tiredness, haemorrhagic manifestations, ulcers in the mouth, recurrent infection	Same as AML. Bone and joint pain are more common. Bleeding as presentation is less common
2. Age	Relatively uncommon in children (common in adults); median age at presentation is 60 years	Mainly a disease of childhood (peak incidence : 3-7 years)
3. Lymphadenopathy and enlarged tonsils	Rare	Common
4. Gum hypertrophy	Seen (specially in M ₅ variety)	Not seen
5. Splenomegaly	Less common (50%)	More common (70%)
6. Chloroma (localised greenish tumour masses in orbit, skin and other tissues)	Common	Usually not seen
7. Anaemia	++	+++
8. Leukaemic meningitis	Less common	More common

* Sternal tenderness is present in both the types of acute leukaemia.

How do you like to manage this case ?

- (A) IMATINIB MESYLATE—It is recommended as first-line treatment in chronic or stable CML, which results in complete 'cytogenetic' response (proved by disappearance of Ph-chromosome) in approximately 76% patients at 18 months of therapy. The complete 'haematologic' response with imatinib is 97% as compared to 69% in patients treated with interferon (IFN)- α and cytarabine at 18 months of therapy. Imatinib inhibits BCR-ABL tyrosine kinase activity and thereby reduces the proliferation of white blood cells. This drug is also indicated in patients presenting in accelerated or blastic phase, and in those who are resistant to IFN- α . The recommended daily dose is 400 mg/day. The side effects encountered are nausea, diarrhoea, skin rash, muscle cramps, fluid retention, periorbital swelling and myelosuppression; in the first year, 3% of the patients may progress to accelerated/blastic phase with imatinib. Newer drugs like nilotinib and dasatinib are now being used in many countries.
- (B) INTERFERON- α —It was the treatment of choice before introduction of imatinib, and patients treated with IFN- α survive longer than patients who are treated with busulphan or hydroxyurea. It is used in doses of 1.5 million unit, subcutaneously on alternate days. It can induce control and maintain the control in chronic phase. There may be elimination of Ph-chromosome in 20% cases. Common side effects are flu-like syndrome, myelosuppression and neurotoxicity (fatigue, depression, myalgia, severe weight loss).
- (C) CHEMOTHERAPY—Hydroxyurea (1-4 g/day) and busulphan (2-4 mg/day) are usually reserved for patients for rapid lowering of WBC, reduction of symptoms and reversal of symptomatic splenomegaly. As cytogenetic remissions with hydroxyurea is uncommon and busulphan produces serious side effects, these drugs are not recommended now-a-days to treat CML. However, hydroxyurea is still being used in many countries.
- (D) SPLENECTOMY (or irradiation of the spleen)—Done to alleviate symptoms of discomfort and pain as a result of repeated splenic infarction, and to correct prolonged thrombocytopenia.
- (E) BLOOD TRANSFUSION—Low platelet count may be raised by repeated fresh blood transfusions.

- (F) BONE MARROW TRANSPLANT (BMT)—Allogenic or syngenic BMT from a matched sibling donor could potentially cure CML. However, in the era of imatinib, the role of BMT in CML is not very clear. Hopefully, long-term results of imatinib will throw some light on it in future.
- (G) Good nutrition, antibiotics for infections and maintenance of proper oral hygiene.
- (H) OTHERS—Oral hydroxycarbamide (used in palliative situations), arsenic trioxide, leukapheresis for symptomatic leucostasis (pulmonary failure or CVA).
- (I) ACCELERATED OR BLASTIC PHASE—
 - (a) Accelerated phase—imatinib, hydroxycarbamide, low-dose cytarabine.
 - (b) Blast crisis—Blastic transformation to ALL responses better than AML. Most patients have a short-lived response to imatinib. However, ultimately BMT may be necessary. Blast crisis have a poor response with all modalities of treatment.

Complications of busulphan :

1. Increased skin pigmentation.
2. Sterility.
3. Dryness of skin and mucous membrane.
4. Pancytopenia (myelosuppression as it acts on early progenitor cells).
5. Interstitial pulmonary fibrosis (busulphan lung) and retroperitoneal fibrosis. Endocardial and marrow fibrosis have been reported.
6. A wasting syndrome similar to that seen in adrenal insufficiency.

* Complications of hydroxyurea are diarrhoea, mucositis or rash.

Features of aplastic anaemia :

It is characterised by hypocellular bone marrow with pancytopenia :

- a) Precipitating factor or the aetiological factor is very often present (drugs, chemicals, radiation).
- b) Severe anaemia and its manifestations—Fatigue, lassitude, vertigo, black-out, tingling and numbness of extremities, palpitation, dyspnoea, chest pain, anorexia.
- c) Leucopenia and its manifestations—Sore throat, fever with chill and rigor due to infection. fatigue, features of depressed immunity.
- d) Thrombocytopenia and its manifestations—Bleeding manifestations into skin as petechiae, purpura, bruise; epistaxis; gum bleeding.
- e) There is absence of splenomegaly, lymphadenopathy, jaundice or sternal tenderness in aplastic anaemia.

Drugs producing agranulocytosis :

1. Chloramphenicol
2. Sulphonamides
3. Chlorpropamide
4. Chlorpromazine
5. Propylthiouracil
6. Cytotoxic drugs
7. Oxyphenbutazone
8. Phenytoin
9. Carbimazole.

*Identification points for **CML** :*

1. Usually an adult person without any suggestive facies.
2. Huge splenomegaly (usually feels hard).
3. Fever (neither double quotidian like kala-azar nor associated with chill and rigor like malaria).
4. Presence of sternal tenderness.
5. Absence of hepato-cellular failure, no jaundice, no lymphadenopathy.
6. Mild anaemia.

Case 25

LYMPHOMA

What is your diagnosis ?

The diagnosis may be told in one of the two ways :

1. It is a case of lymphoma, probably Hodgkin's disease, or
2. It is a patient of generalised lymphadenopathy (for discussion).

Why do you say so ?

This is a patient of lymphoma because of the following :

1. There is presence of fever, malaise, fatiguability, night sweats, loss of body weight and pruritus for last 6 months.
2. Gradual painless swelling of lymph nodes on both sides of the neck, right axilla and both inguinal regions for last 4 months.
3. On examination there are presence of,
 - a) Anaemia (moderate).
 - b) Lymph node enlargement—Cervical, axillary and inguinal nodes are enlarged. The lymph nodes are discrete (not matted), non-tender, rubbery in feel, neither fixed to deeper structures nor to the overlying skin, and without any sinus formation. Local temperature over the lymph nodes are not raised.
 - c) Spleen—Moderately enlarged and non-tender.
 - d) Liver—Moderately enlarged and non-tender.
 - e) Absence of features of superior mediastinal syndrome, bone pain, paraplegia, intestinal obstruction, lump in the abdomen, ascites, pleural effusion at present.
 - f) Absence of sternal tenderness.

Can you specify the type of lymphoma ?

No. Final diagnosis can be given after biopsy of the lymph nodes but the clinical features in this patient suggest that probably it is a case of Hodgkin's disease (see the differentiation between two types).

What is lymphoma?

These are heterogenous group of malignant diseases of lymphoreticular origin (previously, known as reticulosis). The major types are Hodgkin's disease (HD) and non-Hodgkin's lymphoma (NHL).

B-symptoms of lymphoma :

1. Presence of fever above 38°C,
2. Drenching night sweats, and
3. Unexplained loss of more than 10% of body weight within the previous 6 months.

N.B. : 'A' indicates absence of these symptoms. B-symptoms indicate bad prognosis too.

Describe the different patterns of fever in lymphoma :

1. Usually it is a low grade fever unaccompanied by chill and rigor. The fever may be remittent in type like tuberculosis or continued in type resembling enteric fever.
2. Occasionally it is a swinging fever (with ups and downs).
3. Classical 'Pel-Ebstein' fever (10% cases of Hodgkin's disease) is not seen now-a-days. It is a cyclical fever where several days or weeks of fever alternates with afebrile period.

* Pel-Ebstein fever is also noted in brucellosis and hypernephroma.

How to examine the lymphoreticular system ?

This system should be **examined first** in lymphoma. Follow the 'Scheme of examination' in 'Lymphoreticular system'. Ophthalmoscopic examination, though essential, is not in the curriculum of undergraduate students; only mention the heading and write 'not done.'

Method of lymph nodes examination :

Normal adults have approximately 400-450 lymph nodes. In health, usually the lymph nodes are not palpable (inguinal nodes are sometimes palpable) unless it is enlarged. Nodes are palpated symmetrically on **both sides** of the body from above downwards.

(A) LYMPH NODES IN THE NECK—

Method—These lymph nodes are **always palpated from 'behind'** when the patient is sitting or standing with the head bending forward (to relax the muscles in the anterior part of neck).

- a) The upper circular group is palpated symmetrically by both hands (using right hand for right side and left hand for left side) in the following order from front to back :
 - (i) Submental,
 - (ii) Submandibular (under the jaw near the angle),
 - (iii) Tonsillar.
 - (iv) Preauricular,

- (v) Postauricular, and
- (vi) Occipital gland (e.g., sclap infection, pediculosis capitis, rubella).

N. B.:—if one side of the neck is palpated at a time, the neck should be flexed to that side.

- b) Lower horizontal or supraclavicular group of lymph nodes—These are divided into medial, intermediate and lateral groups. Special attention should be paid in palpating left-sided medial group of nodes (**Virchow's**) and **scalene nodes** (present above and behind the head of clavicles, deep to sternomastoids on both sides of neck).
- c) The vertical chain in the middle of the neck—The glands in the (i) anterior triangle, and (ii) posterior triangle are palpated (the triangles are divided by the ipsilateral sternomastoid muscle).

* For palpation of **scalene nodes**, standing behind the sitting patient ask him to shrug his shoulder, and then dip down the palpating index finger behind the clavicle through the clavicular origin of sternomastoid with the patient's neck slightly flexed to that side. Scalene nodes lie in a pad of fat on the surface of scalenus anterior muscle. These nodes are enlarged in bronchogenic carcinoma.

** Few clinicians prefer that glands in the posterior triangle, postauricular and occipital group of nodes are better palpated from front. Occipital nodes are present in the apex of the posterior cervical triangle.

(B) AXILLARY LYMPH NODES—

There are 5 groups. The patient sits on a stool and the examiner sits in front of the patient (only the subscapular group is palpated from behind).

- a) Central group—The left side is palpated by right hand and vice versa for the right side. The patient's arm is abducted and the hand of the examiner is placed in the axilla in such a way that the palm is directed towards the chest. Now the arm is adducted and allowed to rest 'comfortably' (complete relaxation of muscles is necessary) on the examiner's forearm. The other hand of the examiner is placed on the patient's same shoulder. The central group is palpated by sliding the fingers upwards to reach the highest limit of axilla.
- b) Apical group—The same method as described above is applied here but the fingers are pushed as high as possible.
- c) Pectoral (anterior) group—These glands are situated under the anterior axillary fold. For the left axilla, the fingers of right hand are insinuated under the pectoralis major and vice versa for the right axilla. The nodes are palpated with the help of the thumb and fingers.
- d) Brachial (lateral) group—Here the left hand is used for the left side and the right hand for the right side. This group of nodes are palpated against the upper part of the humerus when the examiner's palm directs laterally.
- e) Subscapular (posterior) group—This group lies on the posterior axillary fold. It is palpated from behind when the hand insinuates within the latissimus dorsi, keeping the patient's arm horizontally forward (during palpation examiner's palm looks backwards).

N.B. : Relaxation of muscles is a must during palpation of all the groups of axillary nodes. After examination of one axilla, start examining the other side.

(C) **EPITROCHLEAR LYMPH NODES**—Make the elbow slightly flexed and forearm supinated. The epitrochlear group of nodes are palpated under the thumb if the patient's left elbow is grasped by the examiner's left hand and vice versa for the right. It is better to fix the patient's forearm at the wrist by the examiner's opposite hand. The glands are palpated in the antero-medial region of the lower part of arm (in between the groove of biceps and brachialis muscle) adjacent to the elbow. Both the sides should be examined. Palpable epitrochlear nodes almost always indicate some pathology.

(D) **INGUINAL LYMPH NODES**—This group is commonly enlarged and specially palpable in bare-footed persons due to repeated trauma and infections of the lower limbs. Horizontal chain lies below inguinal ligament and vertical chain lies along saphenous vein. Both the sides are palpated one after another in supine position after extending the thighs.

(E) **POPLITEAL GROUP OF LYMPH NODES**—The patient lies in supine position with semiflexed legs. The popliteal fossa is felt with the fingertips of either hand, when the fingers of both hands being curled into the popliteal fossa and both thumbs rest on tibial tuberosity.

(F) **MEDIASTINAL GROUP OF LYMPH NODES**—It is not possible to physically examine this group of nodes. Their presence can be detected indirectly by percussion of the sternum (read the section on 'Percussion of the sternum or base of the heart'). This group comprises of para tracheal, subcarinal, tracheobronchial and hilar lymph nodes.

(G) **ABDOMINAL LYMPH NODES** (pre- and para-aortic, retroperitoneal)—This group, if enlarged will present as lump in the abdomen, or felt as nodular discrete or matted mass (read the section on Abdominal lump).

* Local examination is incomplete without the examination of the lymph nodes draining the affected area.

** Rudolf Virchow (1821-1902) was a German physiologist, and anatomist of Wurzburg and Berlin.

Description of a lymph node enlargement :

In lymphadenopathy, points to be noted are :

1. Position or anatomical region affected (situation and extent).
2. Number and size.
3. Discrete or matted (margin).
4. Tenderness.
5. Consistency (texture)—Soft (cold abscess), rubbery and elastic (Hodgkin's disease), firm and shotty (syphilis), hard (malignancy).
6. Surface.
7. Mobility—Fixity to skin and surrounding structures (muscles, vessels, nerves or with any viscus) should be examined, i.e., flexed or mobile.
8. Rise of local temperature.
 9. Skin changes [sinus, peau d'orange (orange skin appearance) or any sign of inflammation],
10. Lymphangitis (red, linear, tender lymph vessels radiating proximally)—present or not.
 11. Draining area (e.g., in epitrochlear lymphadenopathy, examine the hand)—it is a must.

* Matted (means periadenitis) nodes are **most commonly found in tuberculosis**, and rarely in chronic lymphadenitis and NHL.

Common causes of cervical lymphadenopathy :

1. Infection (tonsillitis or impacted wisdom tooth), malignancy or ulcer in the oral cavity.
 2. Infection in the ear, eye and nose.
 3. Scalp infection by louse, dandruff.
- * These are common causes of cervical adenopathy which are very often missed.
4. Lymphoma.
 5. Lymphatic leukaemia (acute and chronic).
 6. Miliary tuberculosis.
 7. Metastasis in lymph nodes (from head, neck, breast, thyroid, stomach and lung malignancy).
 8. Infectious mononucleosis, cytomegalovirus infection, HIV infection.
 9. Sarcoidosis.
 10. Rubella infection (postauricular and occipital nodes mainly).

** '**Bull-neck**' — This is classically seen in malignant diphtheria where the space between the mandible and the clavicles bulges with enlarged lymph nodes and oedema.

Causes of axillary and epitrochlear lymphadenopathy :

(A) Axillary adenopathy —

1. Breast carcinoma.
2. Infections of the upper extremity.
3. Lymphoma, melanoma.
4. ALL or CLL.
5. Tuberculosis.
6. Brucellosis.
7. Cat-scratch disease.

(B) Epitrochlear adenopathy —

1. Hand infections.
2. Lymphoma (specially NHL)
3. ALL or CLL.
4. Sarcoidosis.
5. Cat-scratch disease.
6. Tularaemia.
7. Secondary syphilis.

Causes of inguinal lymphadenopathy :

1. Infection, injury or cellulitis of lower limb.
2. Filariasis.
3. Syphilis, tuberculosis, plague.
4. Metastasis from genital malignancy, carcinoma of uterus and rectum.
5. Chancroid, lymphogranuloma venereum (bilateral inguinal 'bubo').
6. Acute lymphoblastic or chronic lymphocytic leukaemia i.e., ALL or CLL.
7. Lymphoma,

Mediastinal, abdominal, auricular and occipital lymphadenopathy :

(A) Mediastinal and hilar adenopathy—

1. Metastasis from bronchogenic carcinoma, breast, stomach, pancreas, colon.

2. Lymphoma (commonly Hodgkin's variety).
 3. Sarcoidosis (hilar glands 1).
 4. Tuberculosis (rare) and fungal disease of lung (rare).
- (B) Abdominal and retroperitoneal adenopathy—
1. Lymphoma.
 2. Carcinoma of stomach, pancreas, colon.
 3. Intestinal tuberculosis.
- (C) Auricular adenopathy—
- a) Preauricular : Conjunctivitis, rodent ulcer, primary oral chancre, herpes zoster ophthalmicus.
 - b) Postauricular : leprosy, rubella.
- (D) Occipital adenopathy—Scalp infection, rubella, secondary syphilis.

What is *Virchow's node* (or *sentinel node ! Ewald's node*) ?

These are medial group of left-sided supraclavicular lymph nodes which lie in between two heads of sternomastoid muscle. They receive lymphatics from,

1. Upper limb (left),
2. Breast (left),
3. Lung (left),
4. Stomach (Troisier's sign), and
5. Testes.

Clinical method for palpation :

Stand behind the patient. Put your right hand over the right side of patient's face and ask him to turn the face to right against your resistance. Two heads of left sternomastoid become prominent and now put your left index finger in between the two heads.

N.B. : Bronchogenic carcinoma commonly produces scalene node enlargement. Supraclavicular nodes (e.g., Virchow's node) are important in relation to malignancy of lung, breast, G. I. tract and genitals.

* **Breast** should be examined for symmetry, ulcers, redness, peau d'orange appearance, dimpling of skin and nipple discharge. Palpate all the quadrants with flat of the hand, and lastly examine the axillary and cervical lymph nodes.

When the enlarged nodes are of pathological significance ?

It is said that lymph nodes are pathologically significant when they are,

1. (Usually) greater than 1 cm in diameter,
2. Firm in consistency,
3. Round in shape,
4. Matted (nodes become adherent), and
5. Tender on palpation.

However, in assessing the significance of enlarged lymph nodes, following factors should always be considered like a) age of the patient, b) the physical characteristics of the nodes, c) anatomical sites involved, and d) the clinical setting.

* Inguinal lymph nodes > 2 cm in size may be considered as 'significant'.

Lymph nodes fixed to surrounding structures :

1. Metastases in the lymph nodes (i.e., malignancy).
2. Tuberculosis.

Painful and tender lymph nodes :

1. Pyogenic infections, diphtheria, infectious mononucleosis, fungal infections.
2. Metastases in the lymph nodes (may or may not be tender).

Lymphadenopathy with sinus formation :

1. Tuberculous lymphadenitis.
2. Actinomycosis.

What is Waldeyer's ring ?

It is the ring of lymphoid tissue within the oral cavity and is composed of,

1. Adenoids,
2. Faucal tonsils, and
3. Lingual tonsils.

What is generalised lymphadenopathy ?

Lymphadenopathy is defined as inflammatory or non-inflammatory enlargement of lymph nodes. 'Generalised lymphadenopathy' is characterised by the involvement of three or more non-contiguous lymph node areas: for example, when the right axilla and both sides of the neck (contiguous affection) are involved, it is not designated as generalised lymphadenopathy. The present case on discussion has involvement of neck, axillary and inguinal lymph nodes, and thus It Is a case of generalised lymphadenopathy.

When a single anatomical area is involved, it Is known as 'localised (regional) lymphadenopathy'.

- * Each side of neck is considered as single lymph node area.

Description of lymph node enlargement in lymphoma :

(A) **HODGKIN'S DISEASE (HD)**—Cervical nodes are Initially affected. Other nodes are involved In course of time and rarely, abdominal and Inguinal nodes are involved.

Description—Enlarged, non-tender with *India-rubbery feel*, smooth surface, not fixed to skin or deeper structures, discrete, no sinus or no rise of temperature in the overlying skin, and there is no softening or any suppuration.

(B) **NON-HODGKIN'S LYMPHOMA (NHL)**—Enlargement of lymph nodes may affect any area initially. There is more chance of involvement of epitrochlear nodes and Waldeyer's ling.

Description—Enlarged, non-lender (rarely tender) with *firm or variegated feel*, smooth or nodular surface, may fix to deeper structures, discrete, without any sinus in the overlying skin, sometimes with elevated local skin temperature; and there is neither softening nor any suppuration.

- * Thomas Hodgkin (1798-1866) was the curator of the museum of Guy's Hospital, London, UK.

Table 18 : Clinical differentiation between HD and NHL

Feature	Hodgkin's disease	Non-Hodgkin's lymphoma
1. Frequency	Less common (30%)	More common (70%)
2. Age	Bimodal peaks seen; larger peak between 15-35 yrs and smaller peak over 50 yrs	Any age; more frequent with increasing age
3. B-svmptoms	Early and prominent	Late and non-prominent (only in 20% cases)
4. Anaemia	Late	Early
5. Dissemination	Unifocal in origin; well localised at diagnosis	Multicentric in origin; widespread at diagnosis
6. Lymph node involvement:		
a! Presentation	90% nodal and 10% extranodal	60% nodal and 40% extranodal
b) Size	Smaller	Larger
c) Rate of growth	Slow	Fast
d) Consistency	Rubbery elastic	Variegated or firm
e) Matting	Rare	May be seen
f) Local temperature	Normal	May be raised
g) Tenderness	Absent	May be present
h) T Epitrochlear nodes	Less common	Common
i) t Waldeyer's ring	Less common	Common
j) t Mediastinal lymph node	More common	Less common
k) T Abdominal lymph nodes	Early involvement of paraaortic nodes	Early involvement of mesenteric nodes
7. Pruritus	Common	Less common
8. Splenomegaly	More common	common
9. G.I. tract involvement	Uncommon	Common
10. CNS involvement	Uncommon	Common
11. Bone marrow involvement	Late	Early
*12 Alcohol test	Common	Uncommon

* After Intake of alcohol, there may be pain in the area of nodal involvement.

** In HD, the disease spreads by contiguity and in NHL, it is non-contiguous (haematogenous) spread.

Pathological classification of lymphoma :

(A) *HODGKIN'S DISEASE* (WHO pathological classification, which is a minor modification of previous Rye classification) :

- Nodular lymphocyte-predominant HD (5%; slowly growing, localised, rarely fatal)
- Classical HD
 - Nodular sclerosis HD (70%)
 - Mixed cellularity HD (20%)
 - Lymphocyte-rich HD (5%)
 - Lymphocyte-depleted HD (rare)

* The prognosis of a), b), c) and d) are respectively a) very good, b) good, c) fair, and d) poor.

** Nodular sclerosis variety is most commonly encountered, followed by mixed cellularity, lymphocyte-rich and lymphocyte-depleted type.

(B) *NON-HODGKIN'S LYMPHOMA* (Rappaport's classification.) :

- a) Nodular—
 - (i) Lymphocytic : poorly differentiated
 - (ii) Mixed: lymphocytic and 'histiocytic'
 - (iii) 'Histiocytic'
- b) Diffuse—
 - (i) Lymphocytic : well differentiated and poorly differentiated
 - (ii) Mixed: lymphocytic and 'histiocytic'
 - (iii) 'Histiocytic'
 - (iv) Lymphoblastic
 - (v) Undifferentiated: Burkitt's and non-Burkitt's lymphoma

N.B. : Lukes and Collins made the most modern classification of NHL according to cytologic types—T cell, B cell, U cell, histiocytic. NHL in 1982 was classified into high-grade, intermediate-grade and low-grade lymphoma (International Working Formulation). Nodular lymphomas are mainly of low-grade (prognostically better) and diffuse lymphomas are mostly of high-grade (prognostically worse).

*** REAL (Revised European-American Lymphoma), a new pathologic classification of NHL, was introduced in 1984, and the new entries over International Working Formulation are mucosa-associated lymphoid tumours (MALT), mantle cell lymphoma and anaplastic large cell lymphoma (Ki-1 lymphoma).

****The classification of NHL is most controversial and has been changed 6 times in past 40 years, like—

1. Rappaport's classification (1966)
2. Lukes and Collins classification (1972)
3. Kiel's classification (1981)
4. International Working Formulation (1982)
5. REAL classification (1984)
6. WHO classification (2001)—most accepted worldwide (based on precursor B, mature B, precursor T and mature T/NK cell)

Describe the spleen in lymphoma :

Spleen has no afferent lymphatics and its involvement indicates vascular invasion even in an apparently localised disease. In 30% patients with normal spleen, Hodgkin's disease was found in the spleen removed at surgery. Again, it is said that splenomegaly does not always indicate involvement by Hodgkin's disease (25% patients with splenomegaly have only reactive hyperplasia). As a whole, spleen is enlarged in 45% patients of Hodgkin's disease. It is of moderate enlargement, firm to hard in consistency, non-tender and at times with irregular surface (previously known as 'hard bake spleen').

In NHL, spleen is often enlarged at the onset of the disease and subsequently may show marked enlargement with features of hypersplenism. *Splenomegaly in NHL usually indicates splenic involvement.* The description of palpable spleen is more or less same in both HD and NHL. Spleen is less commonly involved in NHL than in HD.

Clinical staging (Ann Arbor classification) of your case :

In stage IIIB (as both sides of diaphragm are involved with systemic symptoms).

Ann Arbor staging classification of lymphoma (specially Hodgkin's disease) :

Stage I—Involvement of a single lymph node region (I) or of a single extralymphatic organ or site (IE).

Stage II—Involvement of two or more lymph node regions **on the same side of the diaphragm** (II), or localised affection of an extralymphatic organ or site and of one or more lymph node regions on the same side (above or below) of diaphragm (IIE).

Stage III—Involvement of lymph node regions **on both sides of the diaphragm** (III), which may also be associated with splenic involvement (IIIS) or by localised involvement of an extralymphatic organ or site (HIE) or both (IIISE).

Stage IV—Diffuse or disseminated involvement of one or more extralymphatic organs or tissues, with or without associated lymph node involvement (e.g., involvement of **extralymphatic organ or site** like lung, liver, pleura, bone, bone marrow, skin and subcutaneous tissue).

N.B. : All the four stages are associated with A or B-symptoms. The **lymphatic structures** are lymph nodes, spleen, Waldeyer's ring, thymus, Peyer's patches and appendix. Recently few modifications of this classification has been done (Cotswolds modification). A' (no symptoms) and 'B' (presence of fever, drenching night sweats and weight loss) are added accordingly.

Importance of staging in lymphoma :

Staging is important to establish the extent of the disease and to plan for therapeutic approach. There are two types of staging (i.e., extent of the disease)—clinical stage and pathologic stage. The **clinical** stage (Ann Arbor staging classification) is based on physical examination and different noninvasive tests. The pathologic stage is determined by the informations obtained from invasive tests which include the collection of biopsy specimens from different sites (usually during a staging laparotomy). As NHL frequently disseminate haematogenously and becomes widespread at the time of diagnosis, Ann Arbor staging system has little impact on this variety of lymphoma. One should never forget that the treatment and prognosis in HD depend largely on stage of the disease, whereas In NHL the therapy is usually based on its histological type.

Staging evaluation for NHL is done by :

- | | |
|--|------------------------------------|
| « Physical examination | • Chest X-ray |
| a Presence of B-symptoms | • CT scan (abdomen, pelvis, chest) |
| ® Laboratory tests (CBC, LFT, uric acid, LDH, p ₂ -microglobulin) | • Bone marrow biopsy |
| | • Lumbar puncture |

Confirmation : HD or NHL ?

It is done by biopsy of the lymph nodes. Presence of Reed-Sternberg (R-S) cells confirms the diagnosis of Hodgkin's disease (absence of R-S cells does not exclude Hodgkin's disease).

How to diagnose Reed-Sternberg (R-S) cell or Dorothy Reed cell ?

(A) Features of R-S cell (a giant cell which looks like owl's eye) :

1. Large giant cell with abundance of acidophilic cytoplasm.
2. Mirror image nuclei with vesicular pattern.
3. Chromatin is loosely woven.
4. Multiple nucleoli with perinuclear halo.
5. On immunophenotyping, they are usually CD 15+ and CD 30+.

(B) R-S cells or cells simulating R-S cells are found in:

1. Hodgkin's disease (hallmark of diagnosis)
2. Infectious mononucleosis
3. Breast carcinoma
4. Burkitt's lymphoma
5. Mycosis fungoides
6. Malignant melanoma.

Importance of examination of other systems in lymphoma :

RESPIRATORY SYSTEM—Pleural effusion (often bilateral), collapse of the lung due to mediastinal lymphadenopathy. Always perform the 'mediastinal percussion' and look for the features of superior mediastinal syndrome.

2. CVS—Pericardial effusion.

3. NERVOUS SYSTEM—Cranial nerves involvement, paraplegia, LMN type facial palsy.

G.I. TRACT—Other than hepatosplenomegaly there may be lump abdomen (lymph node lump), jaundice, ascites or signs of intestinal obstruction.

5. BONES—Diffuse bone pain; swelling of bones.

6. SKIN—Skin lesions of lymphoma (lymphoma cutis), signs of generalised pruritus, herpes zoster, erythroderma etc.
7. GENITOURINARY SYSTEM—Haematuria, renal lump, hydronephrosis; occlusion of renal vein may produce nephrotic syndrome; renal failure from ureteric obstruction; testicular mass.

Drugs producing lymphadenopathy (pseudolymphoma) :

1. Phenytoin 2. Cyclosporin A (used in organ transplantation) 3. Carbamazepine 4. Cephaloridine
5. PAS (used in multi-drug resistant tuberculosis) 6. Primidone 7. Hydralazine 8. Allopurinol.

Anaemia in lymphoma :

1. Ineffective erythropoiesis.
2. Haemodilution.
3. Haemolysis (rare).
4. Hypersplenism.

How paraplegia is produced in lymphoma ?

Involvement of thoracic vertebrae is common. It may be produced by :

1. Involvement of body of the vertebra with collapse.
2. Invasion of the epidural space from retroperitoneal lymph nodes with cord compression.
3. Jeopardized vascular supply of the spinal cord.

Jaundice in lymphoma :

1. Hepatic involvement.
2. Hepatitis due to repeated blood transfusion (type B or C).
3. Extrahepatic biliary obstruction (lymph nodes pressing porta hepatis).
4. Haemolysis (autoimmune and rare).

Symptoms relating to G.I. tract :

These symptoms are common in NHL variety. Symptoms may be,

1. Anorexia.
2. Nausea, vomiting.
3. Haematemesis and melaena.
4. Diarrhoea (malabsorption syndrome).
5. Swelling of the abdomen (lump abdomen formed due to adenopathy, or presence of ascites).

Is it possible to have lymphoma without lymphadenopathy ?

Yes, it is possible in abdominal lymphoma.

Possible associations of 'night sweats' in clinical medicine :

1. Tuberculosis.
2. Lymphoma.
3. Chronic myeloid leukaemia.
4. Giant cell arteritis.
5. Brucellosis.
6. AIDS.
7. Amoebic liver abscess.
8. Nocturnal (sleeping) hypoglycaemia.

* **Excessive sweating** is characteristic of hypoglycaemia (drenching sweat), acute myocardial infarction (drenching sweat), thyrotoxicosis, acromegaly, pheochromocytoma (paroxysmal), arecholine sensitivity (present in betel nut) and gustatory hyperhidrosis (autonomic neuropathy).

History with special findings in lymphadenopathy pointing towards a disease :

- fever lymphoma, tuberculosis, ALL, SLE, infectious mononucleosis, AIDS.
- Petechial haemorrhage in palate in a young boy with cervical adenopathy—infectious mononucleosis.
- Hard lump in breast associated with ipsilateral axillary adenopathy—breast carcinoma.
 - « Swelling of feet/legs, non pitting or solid oedema along with inguinal adenopathy—filariasis.
- fever, weight loss, loss of appetite ± night sweats associated with adenopathy—tuberculosis, lymphoma, malignancy, AIDS, At.I.
- Prolonged medication-induced lymphadenopathy—commonly phenytoin-induced (search for gum hypertrophy and acne).
- Lymphadenopathy with skin lesion—SLE (butterfly-rash in face), sarcoidosis (lupus pemo in face), secondary syphilis, amyloidosis.

Causes of generalised lymphadenopathy :

Lymphadenopathy may be an incidental finding. Majority of generalised lymphadenopathy are non-malignant in nature. The causes are :

- | | |
|--|---|
| 1. Lymphoma (HD and NHL). | 8. Collagen vascular disease, e.g., SLE |
| 2. Acute lymphoblastic leukaemia. | 9. Sarcoidosis. |
| 3. Chronic lymphatic leukaemia. | 10. Filariasis. |
| 4. Disseminated or miliary tuberculosis. | 11. Chronic lymphadenitis. |
| 5. Secondary metastasis to lymph nodes. | 12. HIV infection (AIDS). |
| 6. Secondary syphilis. | 13. Amyloidosis. |
| 7. Infectious mononucleosis. | 14. Drug-induced. |

* Brucellosis, blast crisis in CML and Still's disease may give rise to generalised lymphadenopathy.

** Dermatopathic lymphadenopathy—generalised lymphadenopathy from psoriasis or eczema evolving as a reactive process.

Differential diagnosis of generalised lymphadenopathy :

(A) LYMPHOMA—

- Any age.
- Usually cervical group is involved first.
- SVC obstruction.
- Fever, night sweats, loss of weight.
- Hepatosplenomegaly.
- Pleural effusion (often bilateral) or ascites.
- Discrete, rubbery or firm, non-tender lymphadenopathy.

(B) ACUTE LYMPHOBLASTIC LEUKAEMIA (ALL)—

- Children or young adults.
- Fever may be very high.
- Discrete, non-tender adenopathy.
- Moderate splenomegaly with hepatomegaly; severe anaemia.
- Very short course of disease.
- Haemorrhagic manifestations (purpura, gum bleeding, epistaxis), mouth ulcers, sore throat.
- Presence of sternal tenderness.

(C) CHRONIC LYMPHOCYTIC LEUKAEMIA (CLL)—

- Over the age of 45 years (peak 65 years) and M>F; insidious onset.
- Firm, discrete, painless generalised lymphadenopathy.
- Moderate splenomegaly with hepatomegaly.
- Gradual onset of weakness, slowly developing pallor and presence of irregular fever.
- Haemorrhagic manifestations into the skin in late stages.
- Increased frequency of infections.
- Absence of sternal tenderness.

(D) DISSEMINATED OR MILIARY TUBERCULOSIS —

- Children or young adults.
- High rise of temperature with drenching sweats, loss of weight, progressive anaemia, cough and rarely breathlessness.
- Glands are palpable, matted (means periadenitis), usually painless but may be painful, firm, without any rise of local temperature: rarely there may be a sinus present in the skin with formation of cold abscess underneath. Hard and craggy nodes are found in healed and calcified tuberculosis. Upper deep cervical group of nodes are commonly affected.
- Paucity of signs in the chest (miliary). Often few crepitations are heard late in the disease.
- Mild hepatomegaly with mild, tender splenomegaly.
- * Choroidal tubercles may be seen (25%) in the retina by ophthalmoscopy (miliary).
- Mantoux test may be negative, miliary mottlings may be seen in chest X-ray, and caseating granuloma in lymph node biopsy may be found.

* Choroidal tubercles are yellowish and slightly shiny lesions with a diameter of $< \frac{1}{2}$ of optic disc.

(E) SECONDARY METASTASIS IN THE LYMPH NODES—

- Middle or old age commonly.
- Lymph nodes are stony hard or craggy, fixed, non-tender; generally localised, rarely generalised.

3. H/O haemoptysis, melaena, sore throat, breast lump etc.
4. Anaemia with emaciation (cachexia).
5. Presence of primary source like carcinoma of head, neck, thyroid, lung, breast or stomach.

* Though initially non-tender, pain and tenderness of variable intensity may appear lately.

(F) *SECONDARY SYPHILIS*—

1. Usually in young adults.
2. Past H/O exposure and primary syphilis (e.g., genital ulcers).
3. Moderately enlarged, discrete, *firm and shotty*, painless and non-tender adenopathy. Though the distribution is generalised, there is predilection for affection of occipital and epitrochlear nodes.
4. Skin rash, condylomata lata and snail track ulcers (mucous patches) in mouth may be seen.

N.B. : Secondary syphilis is not commonly seen now-a-days? because of the early use of antibiotics in the treatment of 'chancre'. There may be hepatosplenomegaly.

(G) *INFECTIOUS MONONUCLEOSIS*—

1. Children or young adults.
2. Adenitis is mostly cervical (posterior cervical group), may be generalised. Glands are palpable, a bit tender with moderate enlargement.
3. Fever of short duration, malaise, headache, sore throat may be present.
4. Acute onset with a maculopapular rash (like drug rash).
5. Petechial haemorrhages may be seen at the junction of the hard and soft palate.
6. Spleen is often palpable.
7. Atypical lymphocytes in peripheral blood. Positive Paul-Bunnell and Monospot tests.

(H) *SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)*—

1. Young or middle-aged female patients.
2. Present with arthralgia, arthritis, PIJO, myalgia, butterfly-rash in face, oral ulcers or oliguria.
3. Moderate splenomegaly with hepatomegaly.
4. Non-tender, discrete adenopathy.
5. Pleurisy, pleural effusion, pericarditis or renal failure may develop.

(I) *SARCOIDOSIS*—

1. Usually in children or young adults.
2. Generalised adenopathy with frequent involvement of pre- and postauricular, submandibular, epitrochlear and paratracheal nodes.
3. Mild splenomegaly.
4. Classical skin lesions (lupus pernio), uveitis or parotid swelling may be seen. Patient may have cough, dyspnoea, fatigue or presence of erythema nodosum.
5. Bilateral hilar adenopathy in chest X-ray, positive Kveim test, elevated serum angiotensin-converting enzyme (SACE) and negative Mantoux test are characteristic features.

(J) *FILARIASIS*—

1. Any age.
2. Fever with chill and rigor initially with every full moon and new moon. There may be pain felt in the scrotum, breast, vulva, or erythematous and painful cord like swelling in the limbs. Spermatic cord may be thick and tender. Non-pitting oedema of legs with peau d'orange appearance of the skin may be seen.
3. In the chronic stage *elephantiasis* (lymphoedema as well as increased thickness of skin) of scrotum, vulva or legs with pleural effusion (chylous), ascites (chylous) and chyluria may develop.
4. Discrete, non-tender palpable lymph nodes in the inguinal region; may be generalised.

* Peau d'orange appearance and lymphoedema are often associated with filariasis and malignancy.

(K) *CHRONIC LYMPHADENITIS*—

1. Common in cervical and inguinal lymph nodes.
2. Primary focus is usually present in the oral cavity, scalp, genitalia or in the neighbourhood of the affected glands.
3. Moderately enlarged, firm, mildly tender (rarely matted).
4. There may be rise of body temperature.

(L) *HIV INFECTION (AIDS)*—

1. Any age; young adults commonly.
2. H/O homosexuality, promiscuous sex or receiving blood transfusion.
3. Patient may present with infectious mononucleosis-like illness with pyrexia, rash and lymphadenopathy.

4. A subgroup of patients with asymptomatic infection may present with **persistent generalised lymphadenopathy (PGL)** or AIDS-related complex (ARC). Patient with ARC may have chronic diarrhoea, night sweats, fever, weight loss and oral candidosis. PGL is defined as the presence of enlarged lymph nodes (>1cm) in two or more extrainguinal non-contiguous sites for more than 3 months. Lymphadenopathy in AIDS may be due to diseases like tuberculosis or lymphoma.
5. Rapid downhill course.
6. Biopsy of the lymph nodes show reactive hyperplasia.
7. ELISA test for HIV antibody (screening test) and Western Blot test to identify antibodies to specific viral protein (confirmatory test) are positive.

(M) AMYLOIDOSIS—Read the section on 'Hepatosplenomegaly'.

(N) DRUG-INDUCED—See page 230; also known as pseudolymphoma.

How do you like to investigate a case of lymphoma?

1. Blood examination—
 - a) Anaemia (normocytic normochromic).
 - b) Leukaemoid reaction may be seen (common in Hodgkin's disease).
 - c) Mild eosinophilia (10-15% cases) in Hodgkin's disease.
 - d) Lymphocytopenia (bears a bad prognosis)
 - e) High ESR (indicates disease activity).
2. Lymph node biopsy—
 - a) FNAC (fine needle aspiration cytology)—Often not informative. CT-guided biopsy (FNAC) may be performed in abdominal lymphoma. For FNAC, preferred items are : cervical area (inguinal area avoided), discrete node with the largest gland available.
 - b) **Excision (open) biopsy**—'Most important' investigation. **It gives the accurate diagnosis** and helps in evaluation of prognosis (R S cells are hallmark of Hodgkin's disease).
3. Chest X-ray—May show mediastinal widening (HD mainly), pleural effusion or involvement of lung.
4. Ultrasonography or CT scan of abdomen, chest and pelvis—CT scan is the investigation of choice for staging although PET scan is increasingly being used.
5. Pleural aspiration, paracentesis abdominis or lumbar puncture (in paraplegia).
6. Biopsy of pleura, liver or skin lesions.
7. Endoscopy of upper G.I. tract (for haematemesis and melaena) or barium meal X-ray of stomach and duodenum with follow through.
8. Bone marrow examination (aspiration or trephine biopsy)—Helps in the diagnosis and staging of the disease (bone marrow involvement indicates stage IV disease).
9. Bipodal lymphangiography will demonstrate pre- and paraaortic, or retroperitoneal lymph nodes involvement.
10. Liver function tests (may be abnormal with or without hepatic involvement).
11. Biochemical—
 - a) Serum calcium or alkaline phosphatase may be elevated due to involvement of bones.
 - b) Serum copper, LDH and uric acid are often increased in NHL variety.
 - c) Blood sugar, urea and creatinine as a routine investigation.
12. IVP for renal involvement.
13. Skeletal survey by X-ray or Gallium scan (usually osteosclerotic lesion is seen in HD—'ivory vertebra', and osteolytic lesion in NHL variety).
14. Laparotomy (mainly for staging purpose)—rarely required.

* As high-grade NHL may be associated with AIDS, do the HIV testing. For modern classification, send the lymph node biopsy material for immunophenotyping and cytogenetic/molecular analysis.

** In leukaemia, peripheral blood smear and bone marrow examination are more important than lymph node biopsy.

How do you treat a case of lymphoma ?

(A) HODGKIN S DISEASE :

Therapeutic guidelines are,

- a) Indication of radiotherapy—
 - (i) Stage I, and II A with 3 or less areas of involvement.
 - (ii) To lesions causing serious pressure symptoms e.g.. SVC syndrome.

b) Indication of **chemotherapy**—

- (i) All patients with B-symptoms
- (ii) Stage II with > 3 areas of involvement, stage III and stage IV diseases.

Radiotherapy is used in a dose of 3600 to 4400 cGy (daily dose of 200 cGy over 4 weeks). Different modes like involved-field radiotherapy (IF), inverted Y field radiotherapy, mantle-field radiotherapy, extended-field radiotherapy (EF) and total axial lymph node irradiation (TANI) are used.

Regimens of chemotherapy are,

1. MOPP (mustine hydrochloride, vincristine, prednisolone and procarbazine).
2. MVPP (only vinblastine replaces the vincristine of MOPP regimen).
3. ABVD (adriamycin, bleomycin, vinblastine and dacarbazine)—preferred regimen.
4. SCAB (streptozotocin, CCNU, adriamycin and bleomycin).
5. ChIVPP (chlorambucil, vinblastine, prednisolone and procarbazine)—preferred regimen.

* Combined modality (chemotherapy followed by radiotherapy) is applied in bulk disease.

(B) *NON-HODGKIN'S LYMPHOMA* :

Radiotherapy is usually given by involved-field treatment (IF) or total body irradiation (stage I).

Chemotherapy is used in stages II, III, IV in high-grade NHL and in stages IIB, III, IV in low-grade NHL. Regimens of chemotherapy are,

1. Chlorambucil—as single drug oral therapy (in low-grade NHL).
2. CVP (cyclophosphamide, vincristine and prednisolone).
3. CHOP (cyclophosphamide, hydroxyadriamycin, vincristine and prednisolone).
4. BACOP (bleomycin, adriamycin, cyclophosphamide, vincristine and prednisolone).

*** R-CHOP therapy is rituximab combined with CHOP regimen. Addition of rituximab increases the complete response rates and improves overall survival.

N.B. : These drugs are given on a 28-day cycle (one pulse) for a total of 6-8 'pulses' in both HD and NHL. Autologous bone marrow transplantation is a new modality of treatment in NHL. Blood is transfused in severely anaemic patients.

Conclusion :

1. Palpate all the anatomical areas for lymph node enlargement.
2. In a patient with lymphadenopathy, one should be careful about examination of Waldeyer's ring, *breast*, *testes*, non-pitting oedema in legs, sternal tenderness, hepatosplenomegaly, ascites, pleural effusion and tenderness in spine (i.e., paraplegia in a case of lymphoma).
3. All the systems should be examined.
4. In a patient with inguinal lymphadenopathy, examine the legs and sole of the foot for the presence of any ulcer, infection etc.

* A **case of lymphoma** may **be given** as **superior mediastinal** syndrome.

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MISCELLANEOUS**Case 28****CHRONIC KALA-AZAR****What ?*, your diagnosis ?**

The diagnosis may be delivered in one of the two ways :

1. It is a case of huge (sometimes, moderate) splenomegaly with mild (to moderate) hepatomegaly and fever, probably due to chronic kala-azar, or
2. It is a patient of hepatosplenomegaly with pyrexia for discussion (if not properly diagnosed).

Why do you say so ?

This is a case of chronic kala-azar because,

1. The patient comes from an endemic zone (kala-azar is endemic in Bihar, Gangetic belt of West Bengal, along the coast of Brahmaputra in Assam, coastal Orissa and in eastern part of U.P.).
2. Intermittent rise of temperature for last 6 months which occasionally peaks twice in 24 hours.
3. Weakness, malaise, easy fatiguability with loss of body weight (6 kgs over last 6 months) for the same duration in spite of good appetite and persistent fever.
4. Massive non-tender, 'soft' to firm (often with doughy feel) splenomegaly.
5. Moderate enlargement of liver which is firm and non-tender.
6. The ratio of liver and spleen is 2 : 5 or 1 : 2^{*/2}.
7. Presence of moderate anaemia, alopecia and diffuse blackish pigmentation (forehead, perioral and temple region mainly).
8. Received 16 injections of sodium antimony gluconate so far in another city hospital (according to the patient).

Pattern of fever in kala-azar :

1. May be of any type but usually intermittent in nature.
2. To start with, it is of quotidian variety but without definite chill and rigor (mild chill may be present). Fever is usually nocturnal and without signs of toxæmia.
3. Later on, the fever may be of double quotidian (camel hump fever; 20% cases) in nature (i.e., double rise in 24 hours). This type of fever is very specific for kala-azar.
4. The fever is not very high and ranging between 101°-102°F.
5. Chronic cases may be apyrexial.
6. Though the fever is associated with tachycardia, pallor and weight loss, the patient remains ambulatory (in spite of fever).

Table 19 : Relationship of appetite with weight loss

Weight loss + appetite?	Weight loss + appetitei
1. Diabetes mellitus (uncontrolled)	1. Malignancy (advanced)
2. Thyrotoxicosis	2. Infections (tuberculosis, HIV)
3. Chronic kala-azar	3. Depression (endogenous)
4. Malabsorption syndrome	4. Anorexia nervosa
5. Worm infestation in children	5. Addison's disease
6. Pheochromocytoma	6. Chronic renal failure

* T appetite is also characteristic of hypothalamic disorder and bulimia nervosa.

Chief complaints of your patient :

1. Fever and weakness for 6 months, and
2. Heaviness in the left upper abdomen for 2 months.

Other features of chronic kala azar :

1. G. I. Tract—Diarrhoea or dysentery; rarely jaundice in late stage; gingivitis.
2. Respiratory system—Dry cough, chest pain or bronchopneumonia due to superadded infection. Haemoptysis, if complicated by pulmonary tuberculosis.
3. CVS—Palpitation.
4. Haemorrhagic manifestations—Bleeding gum or epistaxis (due to pancytopenia).
5. Skin—Cancrum oris and diffuse blackish pigmentation of the whole body.
6. Reticulo-endothelial system—May present with lymph node swelling (very rare in India).

Physical findings in different systems :

1. General appearance—Dry rough pigmented skin, emaciation, moderate anaemia, scanty hair in scalp, protuberant abdomen, thin legs with occasional oedema feet.
2. G.I. tract—Hepatosplenomegaly, jaundice (in late stage).
3. Respiratory system—Occasionally crepitations are heard in the presence of secondary infection.
4. cvS—Tachycardia, low BP, murmur in the pulmonary area due to anaemia.
5. Haemorrhagic manifestations—Examination of gum and nose is necessary (for gum bleeding and epistaxis).

6. Skin—Increased black pigmentation (so the name kala-azar, which is Hindi version of 'black-sickness') or rarely cancrum oris (ulcerative lesion around mouth).
7. Reticulo-endothelial system—Rarely, there are presence of sternal tenderness (due to severe anaemia) and palpable lymph nodes (very rare in India, found in African form).

What is cancrum oris (noma) ?

This is a form of sloughing ulcerative gingivitis which spread to buccal mucosa, cheek, mandible or maxilla resulting in widespread destruction of bone and soft tissue. It is thought to be due to invasion of tissues by Bacteroids, Fusobacterium and other normal commensals of the mouth. The causes are,

1. Kala-azar (due to neutropenia)
2. Acute leukaemia
3. Measles
4. Enteric fever
5. Immuno-suppression.

Points in the past history you have enquired for :

1. Jaundice (as cirrhosis of liver and thalassaemia may be present with hepatosplenomegaly).
2. Repeated blood transfusions (indicates thalassaemia).
3. Haematemesis or melaena (indicates cirrhosis of liver).
4. Tuberculosis (kala-azar may be complicated by tuberculosis).
5. Swelling anywhere in neck, axilla or groin (indicates lymphoma).
6. I.V drug abuse (may give rise to prolonged fever).
7. Diabetes mellitus and hypertension as a routine enquiry.

Importance of treatment history :

Whether the patient has received any injection for 21-28 days (sodium antimony gluconate) or directly ask the patient if he knows the name of the drugs used so far. Usually a patient of chronic kala-azar is treated by too many doctors with too many drugs.

Describe the splenic enlargement :

It is just like thalassaemic spleen (read the section on Thalassaemia). The difference is that the spleen of chronic kala-azar is 'soft' to firm in consistency and not very firm like thalassaemia. At the end of 1 month of illness, splenic enlargement is about 2-3 cm below the left costal margin and then the spleen enlarges at the rate of 1 cm per month. Spleen may be tender; in splenic infarction. [In case of acute malaria, spleen is usually palpable after second paroxysm of fever (i.e., after 3rd day of illness) and the spleen of enteric fever is palpable after 7 days of illness. 1 The spleen of chronic kala-azar is not so firm like chronic malarial spleen.

The spleen of acute kala-azar (and acute malaria) is always soft in consistency.

Anaemia in kala-azar :

1. Hypoproteinaemia (malnutrition).
2. Bone marrow depression (due to proliferation of parasitised reticulo-endothelial cells).
3. Haemolysis (autoimmune and rare).
4. Hypersplenism (sequestration and splenic pooling).

Causes of jaundice in kala-azar (though rare) :

1. Blood transfusion causing jaundice by hepatitis B or C virus.
2. Portal hypertension with cirrhotic change in liver.

Why there is bleeding kala-azar ?

- X. Before treatment : due to thrombocytopenia.
2. During treatment : with sodium stibogluconate.

How kala-azar is produced ?

- I) Kala-azar is also known as visceral leishmaniasis and is caused by *Leishmania donovani*.
- II) Incubation period averages 1-2 months, but may be several years.
- III) Transmitted by—
 1. Bite of female sand fly (*Phlebotomus argentipes*, the Indian vector).
 2. Blood transfusion (very rare).
 3. Rarely, inoculation of cultures of *Leishmania donovani*.

* The term 'leishmaniasis' encompasses multiple clinical syndromes, i.e., visceral, cutaneous and mucosal leishmaniasis.

What are the differential diagnosis ?

Discuss the differential diagnosis of moderate or huge splenomegaly as your case is. Read the section on 'Hepatosplenomegaly'.

How will you diagnose (chronic) kala-azar?

(A) INDIRECT EVIDENCES :

1. Blood examination—Low Hb with normocytic normochromic anaemia. Leucopenia (neutropenia and eosinopenia) with relative lymphocytosis and monocytosis. Agranulocytosis and thrombocytopenia may occur. *Pancytopenia* is seen. If repeated, there is progressive leucopenia.
2. Aldehyde test (Napier's)—Positive after 3 months of illness. About 1-2 ml of serum is taken and one to two drops of 40% formalin is added to it. A positive result is indicated by 'jellification of milk white opacity' within 20 minutes. This test is falsely positive in multiple myeloma and cirrhosis of liver. Positivity depends upon increase in serum gamma globulin.
3. Antimony test (Chopra's)—Positive after 6 weeks of illness (due to high IgG).
4. Complement fixation test (CFT) with WKK (Witebsky, Klingenstein and Kuhn) antigen becomes positive within 3 weeks. Now-a-days, Kedrowsky's antigen is preferred to WKK antigen.
5. Immunofluorescence test.
6. Haemagglutination test (indirect).
7. Counterimmunoelectrophoresis.
8. ELISA (enzyme-linked immunosorbent assay) test.
9. Others—Direct agglutination test (DAT) with freeze-dried antigen is simple, cheap and a quick test which can be used in the field conditions. Dot-enzyme immunoassay (EIA) and reverse Western blotting also detect antigen. DNA detection by PCR may be done. Another field-tested technique is immunochromatographic dipstick testing of fingerstick blood for antibody to recombinant Leishmanial antigens or synthetic peptides (e.g., rK 39). Detection of leishmanial antigen in urine is being evaluated.
10. Serum electrophoresis—*Hypoalbuminaemia with marked hyperglobulinaemia (IgG)*.

(B) DIRECT EVIDENCES (DEFINITIVE DIAGNOSIS) :

L.D. bodies (Leishman-Donovan bodies) may be demonstrated by,

1. Preparation of buffy coat in blood—The yield is better (83%) if a thick blood film is made or a straight leucocyte edge is produced in the slide, or by using centrifuged citrated blood. The smear is stained by Giemsa's or Romanowsky's stain and examined under oil-emersion lens to see the amastigote form of the parasite containing nucleus and kinetoplast.
2. Splenic puncture or aspiration—Yield is maximum (95-98%), and *gold standard in diagnosis*.
3. Bone marrow aspiration or biopsy—Either taken from sternum or iliac crest (carefully observe for evidence of marrow puncture in your patient)—Yield is 64-85%.
4. Hepatic aspiration (yield is 75-85%).
5. Lymph node biopsy (if palpable) or FNAC—Yield is 64%.
6. Culture in NNN (Novy-MacNeal-Nicolle) medium in a cool incubator at 22°C.
7. Skin biopsy in cases of Post Kala-azar Dermal Leishmaniasis (PKDL).

N.B. : Multiplication of leishmania takes place in macrophages in the liver, spleen and bone marrow. Culture shows promastigote (flagellar) form and stained films show amastigote (non-flagellar) form. The fully developed promastigote form looks like long slender spindle-shaped bodies while the amastigote form is round or oval. Leishmanin skin test is not much informative (negative in acute kala-azar).

HIV patients may be infected with *Leishmania donovani* if ever been in leishmaniasis-endemic areas. The sensitivity of serologic tests is lower in HIV-infected patients than in non-HIV-infected patients.

What is acute kala-azar and how it is diagnosed?

Early kala-azar of 2-6 weeks duration is 'acute kala-azar'. which is diagnosed by,

1. Blood examination—Progressive leucopenia with relative lymphocytosis and monocytosis.
2. Complement fixation test (CFD).
3. Direct agglutination test (detects IgM antibody and a sensitive indicator of acute disease).
4. Counterimmunoelectrophoresis.
5. Demonstration of L.D. bodies from,
 - a) Buffy coat preparation, and
 - b) Bone marrow aspiration or biopsy, from sternum or iliac crest.

N.B. : *Aldehyde test is negative in acute kala-azar.*

Why splenic puncture is not advocated in acute kala-azar?

In acute kala-azar, spleen is not sufficiently enlarged to be punctured.

Complications of kala-azar?

1. Pulmonary tuberculosis or intercurrent infection (pneumococcal pneumonia) in lung.
 2. Cancrum oris.
 3. Pancytopenia with its consequences.
 4. PKDL (post kala-azar dermal leishmaniasis)—skin lesions (macules, papules, nodules, patches) are most prominent in face that develops during or after therapy. D/D of PKDL are syphilis, leprosy and yaws.
 5. Portal hypertension (10%).
 6. Malabsorption syndrome, bacillary dysentery.
 7. Splenic rupture (rarely).
 8. Amyloidosis.
 9. Measles.
 10. Severe epistaxis, malnutrition, uveitis.
- * Death is usually due to bacterial infection or uncontrolled bleeding.

How do you like To treat kala-azar?

1. Sodium antimony gluconate or sodium stibogluconate (pentavalent antimonial)—It is the drug of choice; 1 ml of injection contains 100 mg of the drug. The dose is 20 mg/kg in I.V (preferred) or deep I.M route as single daily dose, for minimally 28 days (maximum daily dose 850 mg). The dose may be repeated after 2-4 weeks for 40-60 days in patients with relapse or incomplete responses.
2. Meglumine antimonate (pentavalent antimonial)—50 mg/kg daily for 15 days (alternative drug).
3. For resistant cases ;
 - (i) Amphotericin B (deoxycholate)—0.5-1 mg/kg, I.V by slow infusion daily or on alternate days (for 20 days). The drug is nephrotoxic.
 - (ii) Pentamidine isethionate—2-4 mg/kg, three times a week for 5-25 weeks depending on the response (usually 15-30 injections are given).
4. Recent advances :
 - (i) AmBisome, the lipid formulation of amphotericin B or liposomal amphotericin B may be used, I.V in resistant kala-azar in a dose of 3 mg/kg/day over a short period of time (given on days 1 to 5, and 14 to 21; may be repeated). This expensive drug takes the advantage of 'target-specific drug delivery system'.
 - (ii) Miltefosine—it is a cytotoxic drug which is used in a daily dose of 2.5 mg/kg/day, orally in two divided doses for 28 days (i.e., 50 mg capsule twice daily). The drug gives excellent results in areas resistant to antimonials, specially in India (particularly in Bihar and North-East parts).
 - (iii) Aminosidine (aminoglycoside paromomycin)—15-20 mg/kg, I.M for 21 days.
 - (iv) Sitamaquine (an 8-aminoquinoline)—being field-tested in various regions and have a narrow therapeutic window.
 - (v) Addition of allopurinol—20-30 mg/kg, daily in three divided doses with sodium antimony gluconate.
 - (vi) Recombinant human interferon- γ (in seriously ill patients or treatment failures) in a dose of 100 pg/m² body surface/day given by S.C route for 28 days along with standard dose of sodium antimony gluconate.
 - (vii) Fluconazole or ketoconazole may be used in a dose of 3 mg/kg daily.
 - (viii) Adjunctive splenectomy is done in some drug-resistant kala-azar.
5. Treatment of anaemia, secondary infections in lung, pulmonary tuberculosis, intercurrent bacterial infections, bleeding manifestations and malnutrition are done accordingly.

* Urea stibamine [discovered by Dr. (Sir) Upendra Nath Brahmachari], the first drug for kala-azar, is not used now. Remember, incomplete therapy of initial illness results in relapse with drug-resistant organisms, which is a burning problem in India.

** First line drugs are pentavalent antimonials and lipid formulation of amphotericin B.

*** **Relapse** (reappearance after cure) : Sodium stibogluconate for 40-60 days (i.e., same dose with doubling the duration. **Resistant** kala-azar (no response to therapy): pentamidine isethionate, liposomal amphotericin B, miltefosine, interferon- γ and ketoconazole.



Left-sided **partial ptosis** with **lateral squint** due to IIIrd cranial nerve palsy (recovering); increased **frowning of forehead** is noted to overcome the drooping of upper eyelid



Motor component of **Vth Cranial nerve paralysis** resulting in wasting of muscles of mastication, eg, temporalis (suprazygomatic portion) and masseter (infrazygomatic portion)



Angular stomatitis due to vitamin B-complex and iron deficiency in a chronic debilitated patient; excoriation and crusting of skin around nostrils are seen



Red-brown flat-topped papules or plaque-like hyperpigmented lesions over both shins in type 2 diabetes mellitus – the **diabetic dermopathy**



Cyanosis of the fingers in a patient of systemic lupus erythematosus – the **Raynaud's phenomenon**; numbness, burning and pain in the fingers are associated with



Digital infarction, painful **digital ulceration**, periungual erythema in **vasculitis** due to microscopic polyangiitis



Classical **palmar erythema** involving thenar and hypothenar eminences, webs and pulps of the fingers in cirrhosis of liver



Bilateral **wrist** and **foot drop** resulting from consumption of adulterated mustard oil

Side effects of different drugs used in kala-azar :

- (A) Antimony preparations—nausea, vomiting, diarrhoea, arthralgia, myalgia, fever, metallic taste, hepatitis, pancreatitis, arrhythmias, Q-Tc prolongation (monitoring should include ECG) or even death; cyanosis, rapid and irregular pulse, dyspnoea and shock are known as 'Nitritoid crisis'; anaphylaxis after 6th or 7th injections.
- (B) Amphotericin B—fever with chills, hypokalaemia, arrhythmias, renal toxicity, myocarditis.
- (C) Pentamidine isethionate—hypoglycaemia, hypotension, tachycardia, irreversible induction of diabetes mellitus and even sudden death.
- (D) Miltefosine—G. I. symptoms like vomiting and diarrhoea, reversible hepatotoxicity and nephrotoxicity. It should be used with caution in females as it is abortifacient and teratogenic in animals.
- (E) Sitamaquine—Methaemoglobinaemia, nephropathy.

Assessment of progress in kala-azar :

It is assessed by, 1. Symptoms 2. Splenic size 3. Hb estimation, and 4. Serum albumin level.

Case 27

DIABETES MELLITUS

What is your diagnosis ?

This is a case of type 2 diabetes mellitus (DM) with evidences of renal involvement (K-W syndrome) and peripheral neuropathy.

What are the chief complaints?

1. Gradual swelling of the whole body for last 6 months, and more for last 2 weeks.
2. Tingling and numbness of the extremities for last 2 years.
3. Dimness of vision for last 4 years.
4. Increased volume with frequency of urine, increased thirst and appetite for last 15 years.

what is the H/O present illness?

This obese male patient aged about 55 years was normal till 15 years back. The onset of the disease was insidious and it was gradually progressive. He was admitted in this hospital 10 days back and has improved slightly with treatment.

All the symptoms regarding increased appetite, thirst and urination started 15 years back. The patient used to drink 3-4 litres of water daily. He had to wake up 3-4 times every night for urination and total urinary output per day was approximately 3.5 litres. There is absence of burning sensation during micturition at present. Though he had a good appetite, he had lost substantial body weight for the last few years. He also complained of increased fatigability and exhaustion even while at rest.

For last 6 months (more for last two weeks), he observed gradual swelling of the whole body which started from the face. The swelling was a bit posture-dependent and pitting in nature. There was no H/O rise of temperature. For the last two weeks, the total daily urine output was approximately 1.5 litres. The patient was admitted in the hospital for these ailments.

For last four years, he had dimness of vision and after using spectacles it was not corrected much. He complained of tingling sensation of the extremities (hands and feet) with weakness of the limbs for last two years. For last fifteen years, he was on oral hypoglycaemic agents (not too regularly) prescribed by different physicians. Once he received inj. insulin for treatment of perianal abscess.

There was no H/O fever, palpitation (thyrotoxicosis), tremor (thyrotoxicosis), haemoptysis (pulmonary tuberculosis), cough (respiratory tract infection), leg pain during walking (large vessel involvement), skin lesions (diabetic dermopathy), chest pain (IHD), impotence or nocturnal diarrhoea (autonomic neuropathy), itching or any foot ulcer. He suffered from repeated attacks of burning micturition during the last fifteen years (UTI). He did not give any H/O unconsciousness (hypoglycaemia, CVA etc.).

* Add past, personal and family history as described below.

** Exclude the causes of substantial weight loss (e.g., thyrotoxicosis).

Importance of past, personal, family and treatment history :**(A) PAST HISTORY :**

1. No H/O tuberculosis (as a complication of DM).

2. No H/O recurrent pain abdomen (occurs in chronic pancreatitis).
3. No other significant serious illness in the past (IHD, lower respiratory tract infection. TIA etc.).
4. All the symptoms started when he was on 40 years (type 2 DM).
5. H/O frequent boils and abscess at various parts of body; delayed wound healing.
6. H/O frequent change of refractory error in eye.

(B) PERSONAL HISTORY :

1. Married with three children (two daughters and one son)—All are well at present.
2. No addiction (non-smoker, non-alcoholic).
3. Sedentary habit; obese.

(C) FAMILY HISTORY:

1. Father was diabetic and died of IHD, 10 years back. Mother is not suffering from DM.
2. Brothers (two) and sisters (one) are healthy and not suffering from DM.
3. No H/O hypertension in the family.

(D) TREATMENT HISTORY :

1. Received glipizide, pioglitazone and metformin in varying combinations for last 15 years; the intake of drugs was not too regular. He was once prescribed inj. insulin.
2. Treated by norfloxacin during the attacks of UTI.
3. Received multivitamins for tingling sensation of the extremities.
4. He had never received corticosteroid and thiazides (may cause secondary diabetes).

Special points in the physical examination of your patient :**(A) GENERAL SURVEY:**

1. Decubitus—Of choice.
 2. Facies—Moon face (puffy with baggy lower eyelids).
 3. Nutrition—Could not be assessed properly due to the presence of oedema.
 4. Thyroid—Not enlarged.
 5. Neck veins—Could not be properly visualised due to the presence of oedema.
 6. Oedema—Pitting; generalised; parietal oedema present.
 7. Skin—Within normal limit.
 8. Pulse Condition of the arterial wall is thickened. All the peripheral pulses are palpable (press the oedema fluid for few seconds for better palpation of pulse and describe all the points in pulse).
 - 9- BP (i) 180/100 mm of Hg—in lying position.
(ii) 170/95 mm of Hg—in standing position.
 10. Respiration—20/mln, thoraco-abdominal in type; other points—within normal limit.
 11. Temperature—Normal at present.
 12. Scrotum Absence of scrotal oedema and hydrocele at present; penis—within normal limit.
 13. Feet—Absence of trophic ulcer; presence of oedema.
 14. Dehydration—Absent (to exclude ketoacidosis).
 15. Height and weight—essential to calculate body mass index (BMI).
- * Mention all the points in general survey.

(B) GENITOURINARY SYSTEM :

1. No renal lump felt.
2. Renal angle—Non-tender.
3. Urinary bladder—Empty (by percussion).
4. Scrotal oedema—Absent.
5. Prepuce, glans penis, anus—within normal limit.
6. Genital discharge per urethra—absent.
- [7. Examine vulva, vagina, and perform per vaginal examination in females]

(C) NERVOUS SYSTEM :

- a) The patient is conscious, alert, oriented and co-operative.
- b) Higher functions with speech—Normal.
- c) Cranium and spine—Within normal limit.
- d) Neck rigidity—Absent.

- e) Cranial nerves—Normal (sometimes, there is 3rd, 4th or 6th nerve palsy in diabetes). Ophthalmoscopy—Not done.
- f) Motor functions—
 - 1. Nutrition—Could not be assessed properly due to the presence of oedema. No foot drop.
 - 2. Tone—A bit diminished in all four limbs.
 - 3. Power—Grade IV in all four limbs.
 - 4. Coordination—Within normal limit.
 - 5. Involuntary movements—None.
- g) Sensory functions—
 - 1. superficial—Pain, touch and temperature sensations are lost over the distal parts of all limbs. Impaired sensation in the proximal part of all limbs and the involvement is rather patchy in nature.
 - 2. Deep—
 - (i) Vibration sense—Lost (mostly over the medial malleoli).
 - (ii) Joint sense—Impaired distally.
 - (iii) Position sense—Impaired distally.
 - (iv) Muscle sense—Calf muscles are tender on palpation and squeezing (Abadie's sign).
 - 3. Cortical—Impaired (in a patient of peripheral neuropathy, cortical sensations can not be tested properly).
- h) Reflexes—
 - 1. Superficial reflex :
 - (i) Abdominal—No response (due to peripheral neuropathy/ascites).
 - (ii) Cremasteric—No response.
 - (iii) Plantar response—No response.

* Superficial reflexes showed 'no response' bilaterally.

2. Deep reflex :	Right	Left
(i) Ankle jerk	Lost	Lost
(ii) Knee jerk	Diminished	Lost
(iii) Biceps jerk	Normal	Normal
(iv) Triceps jerk	Normal	Diminished
(v) Supinator jerk	Normal	Normal
(vi) Clonus	Absent	Absent
3. Visceral reflex :	Within normal limit.	

- i) Trophic changes—Absent (no bed sore, no trophic ulcer in sole of feet). Charcot joint absent,
- j) Cerebellar functions—Normal.

k) Autonomic functions—

- (i) Tachycardia - Absent.
- (ii) Postural hypotension - Absent.
- (iii) Sweating - Normally present all over the body.

1) Gait—Could not be tested.

Romberg's sign — Could not be tested properly (due to anasarca and weakness).

(D) G.I. SYSTEM :

- 1. Teeth, gum, tongue, oral cavity and breath (odour)—within normal limit.
- 2. No hepatosplenomegaly.
- 3. Ascites — Present.
- 4. Parietal oedema in abdomen—Present.
- 5. Venous prominence over abdomen—Absent.

(E) CVS :

- 1. Precordium—No deformity.
- 2. Thrill—Absent.
- 3. Apex—Left 5th ICS, on the left MCL with normal character.
- 4. Heart sounds—Normal.

5. Murmur—Absent.
6. Pericardial rub/opening snap/split/click—Absent.

(F) RESPIRATORY SYSTEM :

1. Respiration—20/min, thoraco-abdominal (due to ascites), regular; absence of air hunger.
2. Trachea—Central in position.
3. Parietal oedema in the chest wall—Present.
4. Percussion—Normal resonant note on both sides.
5. Breath sound—Vesicular in type.
6. Added sounds—Nil.
7. Vocal resonance—Normal on both sides.

(G) LYMPHORETICULAR SYSTEM :

1. Sternal tenderness—Absent.
2. Lymphadenopathy—Absent.

(H) EXAMINATION OF THE SKIN :

Abscess, balano-posthitis, candidosis, xanthoma, xanthelasma or trophic ulcers are absent; diabetic dermopathy is present over shins bilaterally.

(I) EXAMINATION OF THE EYES :

1. No styne.
2. Pseudo Argyll Robertson pupil—Absent.
3. Cataract (early) in both eyes.
4. Glaucoma—Absent.
5. Ophthalmoscopy—Not done.

(J) URINE EXAMINATION DONE AT THE BEDSIDE :

1. Sugar—Present.
2. Protein—Present.

N.B. : Ophthalmoscopy (fundoscopy)—though not required in undergraduate curriculum, it should always be performed in postgraduate examinations.

What is diabetes mellitus ?

It is a clinical syndrome characterised by high blood sugar level (hyperglycaemia) **and glycosuria** due to relative or absolute deficiency of insulin secretion and/or action, or insulin **resistance that leads** to disturbances in carbohydrate, protein, fat metabolism, and water and electrolyte **homeostasis**.

The diagnostic criteria for diabetes mellitus :

- Symptoms of diabetes plus 'random' blood glucose concentration > 200 mg/dl, or
- 'Fasting' plasma glucose > 126 mg/dl, or
- '2h-postload' plasma glucose > 200 mg/dl during an oral GTT.

The new diagnostic criteria for pre-diabetes and diabetes are :

In the new criteria, the categories of FPG are :

FPG <100 mg/dl = Normal fasting glucose

FPG >100 mg/dl and <126 mg/dl = Impaired fasting glucose (IFG)

FPG >126 mg/dl = Provisional diagnosis of diabetes (on more than one occasion)

The corresponding categories when the oral GTT is used, are as follows :

2 hPG <140 mg/dl = Normal glucose tolerance

2 hPG > 140 mg/dl and < 200 mg/dl = Impaired glucose tolerance (IGT)

2 hPG > 200 mg/dl = Provisional diagnosis of diabetes (must be confirmed on a subsequent day)

* FPG = Fasting plasma glucose; 2 hPG = 2-h postload glucose

** The prognostic significance and outcome (i.e., accuracy) are same whether it is the FPG > 126 mg/dl or 2 HPG > 200 mg/dl (i.e., in diabetes). This is why, in clinical practice, the FPG test is now mostly preferred because of ease of administration, convenience, acceptability to patients and its lower cost.

*** 2-h postload glucose is done by taking 75 g of glucose dissolved in 300 ml water.

**** Results are for venous plasma, the whole blood values are lower.

***** 'Fasting' is defined as no calorie intake (i.e., overnight) for at least 8 hours. 'Random' is defined as any time of the day without regard to time since the last meal.

Classification of diabetes mellitus :

Read the modern aetiological classification from 'Bedside Clinics in Medicine, Part II'. In the recent classification, IDDM has been replaced by the term type 1 and NIDDM by type 2 diabetes mellitus.

Who is a potential diabetic ?

These patients usually have no symptoms, and show no abnormality on examination or by GTT but are susceptible to develop diabetes in any time of their life.

1. When one parent is diabetic and the other have a family history of diabetes.
 2. Diabetes present in both the parents.
 3. Mother of a 'Herculean baby' (> 9 lbs).
 4. Non-diabetic member of a monozygotic twin, where the other is diabetic.
- * Patients with IGT(140-199 mg/dl) and / or IFG (100-125 mg/dl) are now regarded as '**pre-diabetes**'.
Latent diabetic* are persons who show impaired GTT under stressful conditions like pregnancy, infections, physical and mental stress or when over-weight.

Characteristics of Type 1 DM :

1. Age of onset is usually below 30 years.
2. Usually wasting is prominent (or lean patients).
3. Abrupt onset with rapid progress.
4. Pancreatic Islet cells (of Langerhans) are almost destroyed. Plasma insulin is low to absent.
5. Polyphagia, polyuria and polydipsia are classically present.
6. Always responsive to insulin.
7. Unresponsive to oral hypoglycaemic agents.
8. Diabetic ketoacidosis occurs very often.
9. Family H/O diabetes mellitus is usually absent.
10. Presence of other autoimmune diseases with autoantibodies.
11. HLA-DR3 or DR4 in > 90%.
12. Disappearance of C-peptide.
13. High mortality, if untreated.

Characteristics of type 2 DM :

1. Usually starts after the age of 30 years.
2. Obese persons (or overweight).
3. Insidious onset with gradual progress.
4. Pancreatic islet cells are not totally destroyed. Plasma insulin is normal to high.
5. Polyphagia, polyuria and polydipsia—Not so classically seen as in type 1 DM.
6. Responsive to oral hypoglycaemic agents.
7. Insulin therapy—Responsive to resistant.
8. Hyperosmolar hyperglycaemic non-ketotic coma occurs very often.
9. Family H/O diabetes mellitus is usually present.
10. Absence of other autoimmune diseases and autoantibodies.
11. No HLA links; 50% concordance in identical twins.
12. C-peptide persists.
13. Low mortality, if untreated.

Causes of polyuria :

Read the section on 'Acute glomerulonephritis'.

Causes of polyphagia :

1. Diabetes mellitus
2. Thyrotoxicosis
3. Obesity
4. Hungry individuals
5. Binge eating (bulimia)
6. Sometimes, in malabsorption syndrome.

Causes of polydipsia :

1. Polyuria
2. Psychogenic polydipsia (compulsive water drinking)
3. Anxiety, tension, phobia
4. Orators
5. Anticholinergic or antidepressant medication
6. After heavy protein meal.

* Pathophysiology of P_3 symptoms : In diabetes, there is hyperglycaemia → glycosuria → osmotic diuresis → **polyuria** → **polydipsia**. Continuous glycosuria leads to neoglucogenesis from protein which results in wasting of muscles (resulting in loss of weight). Non-utilisation of sugar (in the presence of hyperglycaemia) for energy expenditure is responsible for **polyphagia** or excessive hunger. The classic triad of DM is often polyuria, T thirst and weight loss.

Drugs provoking hyperglycaemia :

Corticosteroids, thiazides, diazoxide, dopexamine, protease inhibitor, clozapine, non-selective beta blockers, cyclosporin, pentamidine, nicotinic acid, α-interferon.

How to suspect DM in an outdoor patient?

1. Classical triad of symptoms (polyphagia, polyuria and polydipsia—the P., **symptoms**).
2. Recurrent abscess, boils, carbuncles or fungal infections; recurrent urinary tract infections.
3. Recurrent stye or chalazion.
4. Unexplained emaciation; undue tiredness or fatigue.
5. Delayed wound healing.
6. Frequent change of glasses due to error of refraction.
7. Cataract appearing at an early age.
8. Sudden, isolated IIIrd cranial nerve palsy.
9. Resistant pulmonary tuberculosis.
10. Non-resolving pneumonia.
11. Silent myocardial infarction.
12. Generalised pruritus, balanitis or pruritus vulvae.

Effects of diabetes on pregnancy :

1. Hydramnios 2. Toxaemia of pregnancy 3. Maternal infections 4. Difficult labour 5. Recurrent abortions 6. Postpartum haemorrhage 7. Puerperal sepsis.

Effects of diabetes on the foetus :

1. Prematurity 2. Stillbirth 3. Macrosomia (large baby) 4. Postpartum hypoglycaemia 5. Respiratory distress syndrome 6. Hyperbilirubinaemia 7. Congenital heart diseases, open neural tube defects.

Differential diagnosis (D/D) of your case :

If the history and presentation are so classical, no D/D are required. Otherwise, differentiate among the causes of polyuria, nephrotic syndrome or peripheral neuropathy.

Complications of DM :

The life-history of diabetes mellitus is full of complications. The patient presents to different specialists or superspecialists with different organs/systems involvement. The common cause of death in treated patients are cardiovascular ailments (70%), followed by renal failure (10%) and infections (6%).

(A) ACUTE (EARLY) :

1. **Hypoglycaemia.**
2. **Diabetic ketoacidosis.**
3. Hyperosmolar hyperglycaemic non-ketotic coma.
4. Lactic acidosis (rare).
5. Infections.
6. Acute circulatory failure.

(B) LATE :

1. **Circulatory abnormalities (macro- and microangiopathy).**
2. **Retinopathy (microangiopathy).**
3. **Neuropathy (peripheral and autonomic).**
4. **Nephropathy.**
5. Gastrointestinal disorders.
6. Recurrent infections.
7. Diabetic foot.
8. Dermatological complications.
9. Miscellaneous — Malignant otitis externa, emphysematous cholecystitis, rhinocerebral mucormycosis, hypertriglyceridaemia, platelet aggregate abnormality etc.

Ocular complications of DM ;**(A) NON-RETINOPATHIC :**

1. Recurrent sty and chalazion.
2. Signs of dyslipidaemia (corneal arcus and xanthelasma) may be present.
3. Anterior uveitis (hypopyon).
4. Rubeosis iridis (moth-eaten appearance of iris).
5. Pseudo Argyll Robertson pupil.
6. Frequent change of glasses due to error of refraction.
7. Snow flake cataract (specific of DM)—Rare.
8. Senile cataract—Accelerated senile cataract.
9. Glaucoma.
10. 3rd, 4th and 6th cranial nerve palsy.

(B) RETINOPATHY :**I. Simple, background or non-proliferative—**

- a) Increased capillary permeability.
- b) Capillary closure and dilatation.
- c) Microaneurysms (earliest change observed by ophthalmoscopy and is known as dots).
- d) Arteriovenous shunts.
- e) Venous dilatation.
- f) Haemorrhages (occur in deeper layer of retina and is known as blots').
- g) Hard exudate (due to leakage of protein and lipids from damaged capillaries, and is very characteristic of DM).
- h) Cotton-wool spots (superficial exudates formed due to 'microinfarction' and is usually seen in diabetes with hypertension).

* c) and f) are collectively known as 'dots and blots'.

II. Proliferative —

- a) Neovascularisation (new vessel formation in response to retinal ischaemia).
- b) Pre-retinal haemorrhage (sub-hyaloid).
- c) Retinitis proliferans.
- d) Vitreal haemorrhage.
- e) Retinal detachment.

** Other causes of microaneurysms are systemic hypertension, SLE, hyperviscosity syndrome and Coats disease (rare). *Microaneurysms are the hallmark of diabetic retinopathy.* Neovascularisation is also seen in sickle cell retinopathy, central retinal vein occlusion and Eales' disease.

What is maculopathy ?

When the macula is involved, there is sudden loss of visual acuity. Macula may be involved by,

- 1) Oedema, 2) Haemorrhage, or 3) Exudate (hard).

* Microaneurysm, venous abnormality, haemorrhage or exudate do not interfere with vision unless they are associated with maculopathy. Sudden loss of vision in DM may also be due to vitreal haemorrhage, haemorrhage in iris or retinal detachment.

Neurological complications of DM :**(A) PERIPHERAL NEUROPATHY :**

1. Distal, symmetrical, mixed sensory-motor polyneuropathy (commonest).
2. Asymmetrical, proximal motor neuropathy (diabetic amyotrophy)—commonly affects quadriceps, adductor magnus muscles.
3. Distal, symmetrical, sensory polyneuropathy.
4. Asymmetric mononeuritis multiplex (painful).
5. Acute mononeuropathy (paralysis of 3rd, 4th or 6th cranial nerves, or sudden foot drop and wrist drop).
6. Radiculopathy (usually involves spinal nerves, over the chest wall or abdomen).

(B) AUTONOMIC NEUROPATHY :

1. Hypoglycaemic unresponsiveness (dangerous, due to lack of warning symptoms).

2. Tachycardia (resting), absence of sinus arrhythmia, sudden cardio-respiratory arrest.
3. Postural or orthostatic hypotension (sustained drop in systolic pressure by > 20 mm of Hg or diastolic pressure by > 10 mm of Hg in standing condition than in lying down position—BP should be measured after standing for at least 3 minutes).
4. Sleep apnoea.
5. Gustatory sweating (circumoral sweating during eating), or disturbance of sweating.
6. Gastroparesis diabeticorum (abdominal fullness, vomiting), biliary dyskinesia.
7. Nocturnal and post-prandial diarrhoea.
8. Anhidrosis (loss of sweating).
9. Impotence (erectile dysfunction), loss of libido and absence of testicular sensation.
10. Retrograde ejaculation, hesitancy or precipitancy in urination.
11. Charcot joint; cold feet (due to loss of vasomotor response).
12. Pseudo Argyll Robertson pupil.

* DM never affects brain. Deep-seated, lancinating pain in the lower limbs as a result of peripheral neuropathy is known as diabetic pseudotabes.

Postural (orthostatic) hypotension is commonly due to fluid depletion, autonomic neuropathy, drugs (vasodilators, diuretics) and idiopathic orthostatic hypotension.

*** Causes of **impotence** (erectile dysfunction) in clinical practice are psychological, diabetes mellitus (autonomic neuropathy), atherosclerosis (vascular), pituitary and testicular failure (endocrinopathy), β -blockers and antidepressants (drug-induced), and miscellaneous (CRF, motor neurone disease, prostatectomy or substance abuse).

Other causes of autonomic neuropathy :

Other than diabetes mellitus (commonest cause), they are :

1. G.B. syndrome.
2. Porphyria.
3. Amyloidosis.
4. Uraemia.
5. Peroneal muscular atrophy.
6. Shy-Drager syndrome (orthostatic hypotension with central nervous system disorder).
7. Riley-Day syndrome (familial dysautonomia).
8. AIDS.

Causes of coma in a diabetic :

1. Hypoglycaemia (commonest cause).
2. Diabetic ketoacidosis.
3. Hyperosmolar hyperglycaemic non-ketotic coma.
4. Lactic acidosis (rare).
5. Other causes of coma (may or may not be associated with diabetes) like,
 - a) CVA.
 - b) Renal failure.
 - c) Hepatic coma.
 - d) Drug-induced coma.

* Except No.2, all are causes of non-ketotic coma in a diabetic.

Renal lesions in diabetes (diabetic nephropathy) :

1. Diffuse glomerulosclerosis (commonest lesion).
2. Nodular glomerulosclerosis (K-W lesion).
3. Arterionephrosclerosis.
4. Chronic interstitial nephritis.
5. Pyelonephritis, perinephric abscess.
6. Papillary necrosis.
7. Various tubular lesions like fibrin cap, capsular drop and glycogen in renal tubules.

* The 'sequence of events' in diabetic nephropathy are :

Glomerular hyperperfusion and renal hypertrophy result in increase in GFR within 1 year -> structural

change in glomerulus and mesangium occurs within 5 years with returning of GFR to normal → at 5-10 years some patients develop 'microalbuminuria' (incipient proteinuria) → clinical nephropathy with 'overt proteinuria' develops at 18-20 years → 'end stage renal disease (ESRD)' is reached at 22-25 years.

In DM, recurrent UTI is very common. ACE-inhibitor therapy may delay the onset of nephropathy.

**** Diabetic nephropathy** (microvascular complication) includes all the lesions occurring in the kidneys of patients suffering from DM; No. 1 and 2 may lead to K-W syndrome.

Clinical features due to renal involvement :

1. Oedema.
2. Hypertension.
3. K-W syndrome (nephrotic syndrome).
4. Chronic renal failure.
5. UTI (usually recurrent).
6. Acute renal failure (due to acute papillary necrosis—rare).

Insulin requirement in renal involvement :

Insulin requirement in DM is reduced with advancing renal insufficiency and is due to :

1. Excretion of insulin binding antibodies (with albumin) in the urine.
2. Reduced rate of degradation of insulin.
3. Uraemia tends to impair neoglucogenesis.

* *Daily insulin production in a normal healthy non-obese adult is 25 units.*

Gastrointestinal manifestations in DM. :

1. Fungal infection within the oral cavity (e.g., candidosis).
2. Gastroparesis diabeticorum (gastric atony and stasis).
3. Motility disturbance of gall bladder, oesophagus, stomach and colon.
4. Chronic gastritis.
5. Diabetic enteropathy (diarrhoea).
6. Chronic pancreatitis (steatorrhoea).
7. Fatty liver and increased incidence of pyogenic liver abscess.
8. Acalculous cholecystitis.
9. Periodontitis; rectal and ischiorectal abscess in poor control of blood sugar.

Why there is diarrhoea and steatorrhoea in DM ?

1. Exocrine pancreatic insufficiency,
2. Coexistent coeliac disease,
3. Abnormal bacterial proliferation in intestine, or
4. Autonomic neuropathy ('diabetic diarrhoea').

Skin lesions in diabetes :

1. Recurrent boils, abscess and carbuncles.
2. Balano-posthitis (inflammation of glans penis and prepuce respectively).
3. Vulval moniliasis (pruritus vulvae).
4. Delayed wound healing.
5. Candidosis of skin.
6. Blisters (usually on feet or hands), bullae (bullous diabeticorum), granuloma annulare, vitiligo.
7. *Diabetic dermopathy*—Small rounded brown plaques with a raised border, which is hyperpigmented and scaly; located over anterior tibial surface (shin spots or spotted leg syndrome).
8. Xanthomas (eruptive).
9. Necrobiosis lipoidica diabeticorum (NLD)—Yellowish plaque-like lesion with a brownish border and atrophic centre over the shin.
10. Lipoatrophy and lipohypertrophy (due to insulin injection), or drug rash.
11. Diabetic foot (due to ischaemia and / or neuropathy)—ulcer or gangrene.
12. Dupuytren's contracture.
13. Scleroderma-like skin lesion (tight waxy skin over shoulder and upper back)—scleroderma diabeticorum.

14. Prayer sign—'Cheiroarthropathy' (stiff hand syndrome or limited joint mobility) i.e., mild, fixed curvature of the fingers which makes it impossible to place the hand and fingers flat on a surface.
 15. Acanthosis nigricans in type 2 DM (in case of insulin resistance).
 16. Ulcers (neuropathic or due to arterial disease), specially in legs or sole of feet.
- * Lipoatrophy or lipohypertrophy—localised atrophy or hypertrophy of subcutaneous fat respectively.

Importance of examination of foot in DM :

1. Ulceration, callosity in skin of sole, cracks and fissures, colour of skin (may have cyanotic hue), anhidrosis, clawed-up toes (a feature of neuropathy), digital gangrene, fungal infection in nails and in between toes.
 2. Temperature of the skin (in ischaemia, feet become cold).
 3. Pulsation in arteria dorsalis pedis and posterior tibial artery (\downarrow pulsation in angiopathy).
 4. Ankle jerk—lost in peripheral neuropathy.
 5. Vibration sense—lost early in DM with neuropathy.
 6. Oedema—present in nephropathy.
 7. Foot deformity in Charcot's arthropathy.
- * Diabetic foot ulcer is due to ischaemia (microangiopathy \pm associated atherosclerosis)/neuropathy/both inchaemia and neuropathy \pm secondary infection.

Causes of 'leg ulceration' and their basic investigations :

When the leg ulcers are present on the lower leg, usually it is due to vascular diseases. The site of ulcer on the lower leg often gives an indication of the underlying aetiology. The common causes are :

- (A) Venous disease : Varicose veins, deep venous thrombosis, deep venous obstruction (e.g., from pelvic growth), incompetent valves.
- (B) Arterial diseases : Atherosclerosis, Buerger's disease, vasculitis.
- (C) Small vessel disease : Diabetes mellitus, vasculitis.
- (D) Neuropathy : Diabetes mellitus, leprosy, polyneuropathy due to any cause, syphilis (rare).
- (E) Haemorrhological : Sickle cell disease, hereditary spherocytosis, chronic haemolytic anaemia (e.g., thalassaemia), cryoglobulinaemia, immune complex disease.
- (F) Tumour : Squamous cell carcinoma, Kaposi's sarcoma, melanoma, basal cell carcinoma.
- (G) Trauma : Trophic ulcer, injury, contact dermatitis, self-inflicted, chronic atopic eczema.
- (H) Infections : Tuberculosis, leprosy, syphilis, yaws, mycotic, tropical ulcer.
- (I) Miscellaneous : Pyoderma gangrenosum, osteomyelitis, lymphoedema.

Basic clinical examination : Temperature of the local skin, site of ulcer (venous—lower leg, ankle with pigmentation; arterial—shin, foot; vasculitis—shin, upper leg, and painful; neuropathy—heel, ball of the great toe; painless), peripheral arterial pulses, ankle jerk, vibration sensation, oedema, local cyanotic hue, blood pressure, thickened peripheral nerves.

Basic investigations :

1. Blood—For anaemia, blood dyscrasias; VDRL; sugar, lipid profile, ANF, ANCA.
 2. Urine—For sugar.
 3. Bacterial swab—For detection of pathogens.
 4. Doppler ultrasound of lower extremities (both venous and arterial system).
 5. Ankle : brachial pressure index, or ABPI (in health, the value is ≥ 1.0 in supine position)—ABPI in intermittent claudication is 0.4-0.9; ABPI of < 0.4 is found in critical limb ischaemia.
 6. Venography.
 7. Nerve conduction study.
- * Treatment : stoppage of smoking, control of DM/hypertension/dyslipidaemia, local care, antibiotics in infection, advice of chiropodist, and surgery (balloon dilatation) or amputation in selected cases.

Importance of 'hand examination' in DM :

1. Pulp infection, blisters, digital gangrene, fungal infection in nails and in between fingers (inter-trigo).
2. Cheiroarthropathy or limited joint mobility—the patient is asked to join the hands as if in prayer, and the hands can not be apposed (prayer sign).

3. Thickened, tight, waxy skin on the back of the fingers (sclerodactyly-like).
4. Dupuytren's contracture.
5. Trigger finger (flexor tenosynovitis).
6. Carpal tunnel syndrome is common in diabetics.
7. Small muscle wasting (due to emaciation or peripheral neuropathy).

* **Rheumatological complications** : Cheiroarthropathy, flexor tendinopathy, adhesive capsulitis, osteoarthropathy, Charcot arthropathy and diffuse idiopathic skeletal hyperostosis (DISH).

Circulatory and CVS complications in DM .

1. **Microangiopathy** (responsible for '**triopathy**', i.e., retinopathy, nephropathy and neuropathy).
2. Intermittent claudication due to accelerated atherosclerosis (**macroangiopathy**; macrovascular disease includes coronary artery disease, peripheral vascular disease, cerebrovascular disease, diabetic foot and hypertension).
3. Gangrene (peripheral vascular disease).
4. Impotence (on a vascular basis).
5. Resting tachycardia and postural hypotension (autonomic neuropathy); cardiac failure.
6. Angina pectoris.
7. Silent myocardial infarction.
8. CVA is common in diabetics.
9. Cardiomyopathy.
10. Hypertension (macroangiopathy, or following renal involvement).

* In DM, BP should ideally remain < 130/80 mm of Hg.

Causes of painless (silent) myocardial infarction :

It is known that 15-20% of myocardial infarcts are painless. The common causes are,

1. Diabetes mellitus with autonomic neuropathy.
2. Patient under general anaesthesia.
3. Elderly patients with cerebrovascular disease or dementia.
4. Few patients may not have pain (greater in women).
5. Inability to recognise the symptom by the patient.

Symptoms of hypoglycaemia :

Prolonged hypoglycaemia may produce permanent brain damage. The symptoms (usually blood glucose becomes less than 55 mg/dl) are due to excessive secretion of catecholamines and dysfunction of CNS. Thus they are divided into two groups like 'autonomic' and 'neuroglycopenic' symptoms :

1. Weakness
2. Hunger
3. Sweating (drenching)
4. Tremor (trembling)
5. Palpitation (pounding heart)
6. Dizziness, yawning
7. Headache
8. Clouding of vision
9. Abnormal behaviour (aggression)
10. Confusion, inability to concentrate and incoordination
11. Diplopia
12. Convulsions
13. Coma.

Autonomic — 2, 3, 4, 5; neuroglycopenic — 6, 8, 9, 10, 11, 12, 13; non-specific—1 and 7.

* Remember, autonomic symptoms predominate in rapid onset hypoglycaemia whereas neuroglycopenic symptoms are the chief complaints in gradual onset hypoglycaemia.

** Diurnal hypoglycaemia : nervousness, sweating, hunger and tremor mainly.

Nocturnal hypoglycaemia : night sweats, bad dreams and early morning headache.

*** Hypoglycaemia is traditionally classified into a) Fasting hypoglycaemia (occurs in the presence of diseases like hyperinsulinism, liver disease, renal disease, endocrine causes and drug-induced), and b) Reactive (post-prandial) hypoglycaemia (occurs in response to meals like Dumping syndrome, galactosaemia).

Important points to diagnose hypoglycaemic coma (or D/D with diabetic ketoacidosis) :

Hypoglycaemic coma is always confused with diabetic coma (ketoacidosis). The characteristic features of 'hypoglycaemic coma' are :

1. History of overdose of insulin or oral hypoglycaemic agents, vigorous exercise or missing a meal.
2. Classical symptoms of hypoglycaemia may be present (already described)—Drenching sweat is very important.
3. Sudden onset of unconsciousness.

4.
 - a) Dehydration—Absent (rather pallor plus cold sweat are present).
 - b) Pulse—Normal volume or full pulse.
 - c) BP—Normal or high.
 - d) Air hunger Absent; rather shallow or normal respiration.
 - e) Smell of acetone—Absent.
 - f) Convulsions—May be present.
 - g) **Jerks—May be brisk.**
 - h) **Plantar response—Often bilaterally extensor.**
5. Urine—Absence of sugar and ketone bodies.
6. Blood—Blood sugar is low; HCO_3 level is normal.
7. Lump of sugar (given orally) or infusion of glucose (given in an unconscious patient) helps in regaining consciousness.

* In diabetic ketoacidosis — The patient will have polyuria, polydipsia, anorexia, vomiting and abdominal pain. Dehydration +; pulse is usually weak, BP is low with dry skin and tongue; air hunger is present with smell of acetone in the breath (Kussmaul's breathing); deep reflexes are dull with flexor plantar response. Sugar and ketone bodies are present in urine; high blood sugar with low HCO_3 level.

Bedside clinical tests for autonomic neuropathy :

There are battery of tests to diagnose autonomic neuropathy but the commonly practised cardiovascular reflexes are described below. In autonomic neuropathy, there will be :

1. Response of Valsalva manoeuvre on R-R interval in ECG (absence of T in R-R interval).
2. Change of heart beat and BP, in response to change of posture (absence of slight T in pulse and BP on standing).
3. Blood pressure response to sustained handgrip (absence of normal T in BP during handgripping).
4. Heart rate response to deep breathing (absence of **T** heart rate on deep inspiration).
5. Resting tachycardia (i.e., tachycardia at rest).

* Simply one can examine at the bedside for light reflex, accommodation reflex, pupillary resistance to mydriatics, or sweating.

What are the causes of sudden death in diabetes ?

Probably it is due to autonomic neuropathy and the different hypothesis put forward are :

- a) Patients do not respond normally to hypoxia and sustain cardio-respiratory arrest.
- b) Apnoea.
- c) Cardiac arrhythmias.

Investigations you like to perform in a case of DM :

1. Blood—
 - a) Sugar (fasting and PP; for diagnosis and monitoring) and glycosylated Hb (HbA or HbA_{1c} — comprising 4-6% of total Hb and reflects blood glucose concentration for preceding 6-8 weeks).
 - b) TC, DC and ESR; triglycerides, LDL, VLDL, HDL and cholesterol (lipid profile); urea and creatinine; uric acid.
2. Urine (R/E)—with special reference to specific gravity, RBC. pus cells, protein, sugar, ketone bodies, casts. Culture and sensitivity tests are done, and 24-hours protein excretion is measured, if indicated (i.e., in nephrotic syndrome developing as a complication).
3. EGG—To diagnose IHD or to observe the changes due to hypertension (associated).
4. Chest X-ray—Pulmonary tuberculosis and fungal infection of lung are very common in diabetics. Cardiomegaly from hypertension should be looked for. Straight X-ray of abdomen is done to exclude pancreatic calcification (fibro-calculous pancreatic **disease**—commonly seen in T **DM**).
5. Ophthalmoscopic examination (fundoscopy)—for diabetic retinopathy.
6. Barium meal examination of G.I. tract and endoscopy are done to study motility of the G.I. tract.
7. Nerve conduction study (in peripheral neuropathy).
8. Doppler studies of peripheral vessels, if indicated.

* Glycosylated haemoglobin is a parameter for assessing long-term glycaemic control, and for tight control of DM it should be kept below 7%. ESR may be high in the presence of infection, commonly tuberculosis. Glycated (glycosylated) fructosamine, which measures the amount of glycated serum protein (mainly albumin) gives an indication of glycaemic control for over the last 2-3 weeks.

How to classify oral hypoglycaemic agents (OHA) ?

(A) SULPHONYLUREAS :

- a) 1st generation—Chlorpropamide, tolbutamide, tolazamide, acetohexamide.
- b) 2nd generation—Glimepiride, Glipizide, gliclazide, glibenclamide, glyburide.

(B) NON-SULPHONYLUREA AGENTS :

- a) Biguanides : Phenformin (not used now-a-days), metformin, buformin.
- b) Meglitinide analogues (prandial glucose regulators) : Repaglinide, nateglinide.
- c) Alpha-glucosidase inhibitors : Acarbose, voglibose, miglitol.
- d) Thiazolidinediones (also known as 'glitazones' or PPAR γ agonist (peroxisome proliferator-activated receptor- γ)). : Troglitazone (no longer used), ciglitazone, pioglitazone, rosiglitazone.
- e) Dipeptidyl peptidase-4 (DPP-4) inhibitors : Sitagliptin, vildagliptin.

* The OHAs are divided into three categories :

1. Insulin sensitizers : metformin, pioglitazone, rosiglitazone.
2. Insulin secretagogues : glimepiride, glipizide, gliclazide, tolbutamide, chlorpropamide, glibenclamide, meglitinide analogues.
3. Inhibitors of glucose absorption : alpha-glucosidase inhibitors, guar gum

** Parenteral hypoglycaemic agents are :

- Insulin
 - Glucagon-like peptide (GLP-1) analogue—Incretin mimetics like exenatide and liraglutide.
- Amylin agonist—Pramlintide.

How will you correct the fluid loss in ketoacidosis ?

In an established case, the patient usually suffers from a fluid deficit of 7-8 litres. It is advisable to correct the fluid loss in this way :

Normal saline volume	Time
1 litre	in 30 min
1 litre	in next 1 hour
1 litre	in next 2 hours
1 litre	in next 4 hours
4 litres	in next 16 hours

Fluid of choice is normal saline until the blood sugar falls below 250 mg/dl when it may be changed to 5% glucose.

Dawn phenomenon :

When DM patients (type 1 DM mainly) are closely monitored during insulin therapy, it is seen that hyperglycaemia develops in between 3-7 AM. This increase in the insulin requirement is known as Dawn phenomenon and is due to increase in the counterregulatory hormones like cortisol, growth hormone, glucagon and catecholamines. During sleep, there is a surge in the secretion of these hormones. Treatment is aimed at to **increase** the overnight insulin dosage.

Somogyi phenomenon :

This is a manifestation of early morning rebound hyperglycaemia following, possibly unrecognised, hypoglycaemia. Often it is seen that the previous night time dose of insulin was in excess. Treatment is aimed at to **decrease** the overnight insulin dosage. This is commonly seen in children.

* Early morning (3 AM) blood sampling is necessary to differentiate both the conditions. In Dawn phenomenon, both the 3 AM and morning fasting blood show hyperglycaemia. In Somogyi phenomenon, 3 AM blood sample shows hypoglycaemia and morning fasting blood documents hyperglycaemia.

Chronic complications in type 1 versus type 2 DM (with approximate incidence) :

	Type 1	Type 2
1. Retinopathy	90% (after 20 yrs)	15-20% (after 15 yrs)
2. Nephropathy	30-40%	3-16%
3. Neuropathy	60% (after 30 yrs)	60% (after 30 yrs)
4. Ischaemic heart disease	Increases after 30 yrs	2-3 times of normal
5. Peripheral vascular disease	30%	45% (after 20 yrs)
6. Hypertension	50% (after 30 yrs)	50%

* The cumulative risk of nephropathy in type 2 DM varies with ethnic origin (may be 50%).

** Though microvascular diseases are the main cause of death in type 1, many patients of type 2 succumb to macrovascular disease prior to development of significant microvascular disease. Advanced age, development of atherosclerosis as a result of co-morbid conditions like physical inactivity, smoking, hypertension and hyperlipoproteinaemia are important causes of death in type 2 diabetes.

*** Majority of diabetics (90-95%) in India suffer from type 2 DM.

**** In x_1 DM, microalbuminuria is an early predictor of nephropathy while in T_2 DM, it usually predicts future development of malignant angiopathy and cardiovascular complications.

Define metabolic syndrome or 'syndrome X' :

The **NCEP ATP III criteria** goes like this—

Presence of three or more among five of the following :

- High blood pressure > 130/85 mm of Hg
- Hypertriglyceridaemia > 150 mg/dl
- Low HDL cholesterol < 40 mg/dl in men and < 50 mg/dl in women
- Abdominal obesity : waist circumference > 102 cm in men and > 88 cm in women
- High fasting blood glucose > 110 mg/dl

* NCEP = National Cholesterol Education Programme; ATP = Adult Treatment Panel

The International Diabetes Federation (IDF) criteria (needs large waist + any other 2 features) are

- High blood pressure > 130/85 mm of Hg
- Hypertriglyceridemia > 150 mg/dl
- Low HDL cholesterol < 40 mg/dl in men and < 50 mg/dl in women
- Waist circumference > 94 cm in men and > 80 cm in women
- High fasting glucose > 100 mg/dl.

N.B. : WHO criteria of metabolic syndrome requires insulin resistance to fulfil the criteria. '**Insulin resistance**' is clinically suspected by the presence of acanthosis nigricans, skin tags (acrochordons), central obesity, lipodystrophy and features of polycystic ovarian disease.

Outline of management of your patient (T_2 DM) :

1. Diabetic diet.
2. Insulin (insulin requirement is low as there is renal involvement) Used to improve the glycaemic
3. Oral hypoglycaemic agents (reduction or modification of dosage may be necessary in azotaemia).
4. Treatment of hypertension, obesity, dyslipidaemia and coronary heart disease..
5. Treatment of oedema.
6. Management of nephrotic syndrome. Even dialysis may be needed, if azotaemia develops.
7. Control of muscle pain (due to neuropathy) by :
Phenytoin, carbamazepine, pregabalin, codeine or tricyclic antidepressants.
8. Control of UTI and skin lesions, if develops.
9. Care of the feet to prevent 'diabetic foot' (consult cheiropodist).

Conclusion :

1. Examine all the systems in a patient of DM.
2. Palpate all the peripheral pulses (specially pedal pulses). Measure BP both in lying down and upright position.
3. Search for trophic ulcer in foot.
4. Give attention to retinopathy, nephropathy and neuropathy.
5. Do not forget to test the urine sample for sugar and protein, if possible.

CHAPTER II : SHORT CASES

Case 28

CYANOTIC CONGENITAL HEART DISEASES

Clinical presentation :

A young male with H/O anoxic spells, breathlessness and syncope is presented. On examination, it reveals central cyanosis, polycythemia, clubbing, tachypnoea and an ejection systolic murmur in the pulmonary area.

The most probable provisional diagnosis is Fallot's tetralogy.

How do you classify congenital heart disease ?

(A) Cyanotic:

With increased pulmonary blood flow-

- Transposition of great vessels
- Truncus arteriosus
- Total anomalous pulmonary venous connection (TAPVC)
- Double-outlet right ventricle
- Hypoplastic left heart

Normal or decreased pulmonary blood flow-

- Tetralogy of Fallot (TOF)
- Tricuspid atresia
- Ebstein's anomaly
- Pulmonary atresia
- Single ventricle

(B) Acyanotic :

With shunt (le/t-to-right)-

- Atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA)
- Partial anomalous pulmonary venous connection
- Rupture of sinus of Valsalva
- Coronary arteriovenous fistula
- Aortopulmonary window

Without shunt-

- Coarctation of aorta
- Congenital aortic stenosis
- Congenital pulmonary stenosis
- Left heart malformation (congenital mitral stenosis, congenital mitral regurgitation)
- Idiopathic dilatation of the pulmonary trunk

* Common cyanotic congenital heart diseases include 5 T: Tetralogy of Fallot Tricuspid atresia Transposition of great vessels, TAPVC and Truncus arteriosus.

** Commonest congenital cardiac anomaly is bicuspid aortic valve (affecting 1-2% of the population) which goes unnoticed until later in life when the valve is stenosed due to fibrosis or calcium deposition or results in significant aortic regurgitation. VSD is the commonest congenital heart disease.

Components of tetralogy of Fallot (TOF) :

It is the commonest cyanotic congenital heart disease in children above the age of 1 year i.e. who do survive infancy and is commonest among adults. A baby who is cyanotic at birth is unlikely to have TOF. It has 4 distinct abnormalities:

1. Ventricular septal defect (large, non-restrictive, infracristal) - VSD,
2. Right ventricular outflow tract obstruction - infundibular pulmonary stenosis (PS),

3. Overriding of aorta (aorta arises from both left and right ventricles), and
4. Right ventricular hypertrophy - RVH.

Give a brief account of tetralogy of Fallot :

Children with TOF may present with H/O dyspnoea, fatigue, deep cyanosis, hypoxic spells (see below), squatting, syncope (due to severe right ventricular outflow tract obstruction resulting in increased right-to-left shunt with admixture of venous and arterial blood leading to reduced cerebral oxygenation), recurrent respiratory tract infection, haemoptysis or impaired growth. Physical examination reveals prominent a-wave in neck veins, central cyanosis (commonest clinical manifestation and depends on the severity of PS; the more severe the PS is, the more is the right-to-left shunt through V. Cyanosis is also due to mixing of blood in overridden aorta), clubbing, polycythemia (secondary to chronic hypoxaemia), quiet precordium, normal sized heart (no cardiomegaly), mild parasternal impulse (due to VSD there is absence of heave), systolic thrill (rarely felt) and ejection systolic murmur (due to PS) along the left sternal border and in the pulmonary area. *The intensity and duration of the systolic murmur are inversely proportional to the severity of PS.* S₁ is normal, and S₂ is single, loud and represents aortic valve closure (A). Continuous murmurs of bronchial collaterals may be heard over the back. As the VSD is large, the systolic pressure difference between two ventricles remains very low, and thus the VSD murmur remains inconspicuous (silent). The right ventricle is effectively decompressed by VSD. and this is why CCF does not occur in TOF unless complicated by infective endocarditis or anaemia.

The patient may be complicated by syncope (hypoxic spell), paradoxical embolism with brain abscess, infective endocarditis, and complications following marked secondary polycythemia, e.g. intravascular thrombosis developing at different sites. Chest X-ray (PA view) shows normal-sized heart with a characteristic appearance known as 'Coeur en Sabot' (boot-shaped heart with an upturned apex) with oligoemic lung fields. ECG and cardiac catheterization are also done, while echocardiography is diagnostic.

* Mild case—loud and prolonged murmur, severe case—reduced murmur.

Common associations with tetralogy of Fallot :

Other cardiac anomalies associated with TOF are:

1. Patent foramen ovale (present in all).
2. Right-sided aortic arch (25%).
3. Persistent left-sided superior vena cava (10%).
4. Aortic regurgitation.
5. PDA.
6. ASD.
7. Anomalous origin of coronary arteries.
8. Absent left pulmonary artery.

Other connotations regarding Fallot's type of defects :

- Fallot's triology - PS + reversed interatrial shunt (ASD) + RVH.
- Fallot's pentalogy - Fallot's tetralogy + ostium secundum ASD.
- Acyanotic (pink) Fallot's - Where right ventricular outflow tract obstruction (PS) is mild, i.e., mild Fallot's.
- Acquired Fallot's - In 5% cases of VSD, jet of blood comes to infundibulum and results in its hypertrophy, and thus the right ventricle is enlarged.

Complications of TOF:

1. Anoxic spells, syncope, seizure.
2. Paradoxical embolism (thrombus from systemic veins may enter into arterial system through communication between ventricles and results in CVA or infective endocarditis).
3. Infective endocarditis (common).
4. Polycythemia (may produce CVA), cerebral thrombosis, cerebral abscess.
5. Proneness to pulmonary tuberculosis.
6. Growth retardation.

What is hypoxic, anoxic or cyanotic spells?

The child hyperventilates, suddenly becomes more cyanosed and may develop apnoea, syncope, seizures or even sudden death (thus, potentially fatal), specially during waking up, feeding, crying, pyrexia or exercise. Cyanotic spells (Fallot's spells) are common between 2 months and 2 years of age.

Systemic vasodilatation follows feeding, crying etc resulting in an increased right-to-left shunting of blood through VSD with associated decreased pulmonary blood flow. The child adopts *squatting* posture, i.e., sits with flexed hips and knees (knee-chest position) to reduce the cyanotic spells, and thus peripheral systemic vascular resistance is increased which reduces right-to-left shunting with improvement in arterial oxygen saturation and improved cerebral oxygenation; squatting also increases venous return to the heart and thereby increasing pulmonary blood flow. Squatting following physical exertion is almost always pathognomonic of TOF. The hypoxic spells may vary from once in a fortnight / month to multiple attacks in a day.

Management of TOF :

Medical: Iron supplement for anaemia and antibiotic prophylaxis for infective endocarditis are essential. Cyanotic spells are managed by,

1. Squatting or knee-chest position.
2. Humidified 100% oxygen.
3. Morphine (0.1-0.2 mg/kg, I.V).
4. Propranolol (0.1 mg/kg, I.V).
5. Methoxamine (vasopressor) drip.
6. IV infusion of sodium bicarbonate (2 ml/kg) to combat acidosis.
7. If medical therapy is ineffective, emergency surgical shunts may be considered.

Surgical:

1. Blalock-Taussig shunt (preferred operation) - Left subclavian artery-pulmonary artery anastomosis.
2. Waterston-Cooley shunt - Ascending aorta-right pulmonary artery anastomosis.
3. Pott's shunt - Descending aorta-left pulmonary artery anastomosis (not used now).

What is atrial septal defect (ASD)?

It is the most common congenital heart disease in adults and 2-3 times more common in females. There are two main types of ASD, *ostium secundum* (90%, commonest type; at the level of fossa ovalis) and *ostium primum* (5%, rare; antero-inferior to foramen ovale). The third type of ASD is sinus venosus type (5%, posterior to foramen ovale). ASD is an acyanotic congenital heart disease where there is a defect in the atrial septum with shunting of blood from left atrium to right atrium. ASD should never be confused with patent foramen ovale (PFO). PFO is a normal variant, usually remains asymptomatic and not a true septal defect. In ASD, the right atrium not only receives blood from SVC and IVC but also from the left atrium, and thus enlarged in size.

Most of the children are asymptomatic but may complain of weakness, dyspnoea (effort intolerance), palpitations (due to atrial arrhythmia); the patients are prone to respiratory infections. The pulmonary vascular resistance which is low or normal at childhood goes high up as the patient reaches third or fourth decade, giving rise to pulmonary hypertension. As pulmonary hypertension develops, the initial left-to-right shunt reverses to right-to-left shunt (Eisenmenger syndrome). The patient may develop right heart failure or atrial fibrillation in the adult life.

* In ASD, atrial fibrillation is common, and infective endocarditis is rare (due to slow flow of blood).

Associated diseases / anomalies with ASD :

1. Down's syndrome (ostium primum).
2. Marfan's syndrome (ostium secundum).
3. Partial anomalous pulmonary venous connection.
4. Persistent left-sided superior vena cava.
5. Pulmonary stenosis.
6. Lutembacher's syndrome (ASD with rheumatic mitral stenosis).
7. Mitral valve prolapse or floppy-valve syndrome (mitral regurgitation).
8. Holt-Oram syndrome (upper limb dysplasia, hypoplastic thumb, fingerisation of thumb, absent radius, accessory nipple, pectus excavatum).

Basic physical findings in ASD :

1. ASD may have a systolic thrill (10%) in the pulmonary area. If the thrill is very prominent, think of associated PS.
2. Prominent left parasternal lift.
3. Wide and fixed splitting of S₂ (hallmark of diagnosis) with a loud P₂. 'Fixed' means that the splitting does not varies with the phases of respiration.

4. Mid-systolic pulmonary ejection murmur (grade II or III) due to increased flow across the normal-sized pulmonary valve, which is best audible at left 2nd or 3rd intercostal space. No murmur is produced across the septal defect due to low pressure gradient between two atria.
5. A mid-diastolic flow murmur across the tricuspid valve and right-sided S₂, are audible in lower left parasternal region.
6. If pulmonary hypertension develops, few of the auscultatory findings change (Eisenmenger's syndrome) - see below.

* Pulmonary stenosis (PS) is the closest D/D of ASD. PS will have systolic thrill in the pulmonary area, soft P and left parasternal heave.

Investigations performed in ASD :

Chest X-ray - It shows a prominent pulmonary artery (a big pulmonary artery segment is seen and can be traced upto diaphragm; end-on view of juicy pulmonary artery is seen as white dots) and pulmonary plethora. ASD also reveals hypoplastic aortic knuckle, mild to moderate cardiomegaly, prominent right atrium and right ventricle.

ECG - Usually right axis deviation (85%); left axis deviation in ostium primum, and 10% having normal axis. RBBB is seen, which is partial or complete. Complete heart block may be revealed in ostium primum type; large ASD may have prolongation of P-R interval.

Cardiac catheterization is performed while echocardiography is diagnostic, which demonstrates the defect and the size of the chambers.

Management of ASD :

Medical : Treatment of respiratory infections, arrhythmias, heart failure and infective endocarditis (though rare).

Surgical : Upto 2 years of age, ASD can close spontaneously. However, children in the age group of 3-5 years without complications and having a significant shunt (i.e., pulmonary to systemic flow is > 1.5:1) are best candidates for surgery. The defect is repaired by Dacron patch or direct suturing.

What are the characteristics of ventricular septal defect (VSD)?

Excluding bicuspid aortic valve, VSD is the commonest cardiac malformation occurring once in approximately 500 live births or accounting for 30% of all congenital cardiac defects, either in isolation or with a range of other malformations. VSD is an acyanotic congenital heart disease where there is a defect in the ventricular septum with shunting of oxygenated blood from high-pressure left ventricle to low-pressure right ventricle. When pulmonary blood flow becomes very large and pulmonary arterial pressure increases, there is reversed shunting of blood from right-to-left (Eisenmenger's complex) with development of central cyanosis. The septal defects may vary in size, shape and number, and can be either in membranous part or in muscular portion of the ventricular septum; perimembranous defects are most common (80%), and may be supracristal (subpulmonic) or infracristal (subaortic or behind the tricuspid valve).

Basic physical findings in VSD :

The apex beat is hyperdynamic. A systolic thrill may be palpable at the left parasternal region. Left parasternal lift or heave may be felt. The S₁ and S₂ are usually masked by the murmur. The murmur is typically rough and pansystolic / holosystolic, best heard in left 3rd and 4th intercostal space close to the sternum but radiating all over the precordium. A mid diastolic flow murmur across the mitral valve may be audible at the apex in large VSD.

Small VSD ('maladie de Roger') - Very often found in routine examination. It is the asymptomatic, small haemodynamically insignificant VSD where thrill and pansystolic murmurs are very prominent. Majority (90%) closes spontaneously by the age of 10 years.

Moderate VSD - The patient comes with fatigue, dyspnoea or pulmonary infections. Cardiac size is enlarged (LVH). Palpable systolic thrill and pansystolic murmur radiated towards right chest are prominent.

Large VSD - Usually develops CCF at 1 year; at 9 - 10 years of age, eventually Eisenmenger's complex develops. There is cardiac enlargement. Usually, there is no thrill and murmur is not so rough.

* Signs of closing VSD - Holosystolic murmur gets shorter, softer and gradually disappear.

Chest X-ray reveals enlargement of LA and LV along with pulmonary plethora. **ECG** features biventricular hypertrophy in larger defects. A small VSD shows no abnormality in chest X-ray or in ECG. **Echocardiography** identifies the site, size and haemodynamic consequences of VSD.

*** **Management** - Medical treatment is done against infective endocarditis and CCF, Surgical closure of moderate and large VSDs (by Dacron patch or direct suturing) is ideally performed before 2 years of age or before the development of severe pulmonary hypertension.

What is patent ductus arteriosus (PDA) ?

The ductus arteriosus connects the **left pulmonary artery** just after the bifurcation of the main pulmonary artery with the **descending aorta** just distal to the origin of left subclavian artery. In the fetus, the channel (ductus arteriosus) diverts blood from the pulmonary to systemic circulation. Physiological closure occurs soon after birth (due to high oxygen concentration in lungs and reduced pulmonary vascular resistance), and anatomical closure is completed within 1-2 weeks after birth. In a malformed duct with less elastic tissue, the patient may live with patent ductus arteriosus (PDA). As pressure in aorta is higher than pulmonary artery, blood shunts in PDA from aorta-to-pulmonary artery (left-to-right). Thus, venous return to left heart is increased resulting in left ventricular volume overload and ultimately its failure. Over the time, pulmonary hypertension with reversed shunting (right-to-left), i.e., Eisenmenger's syndrome develops.

PDA is common in females, premature babies, babies born at high altitudes or in continual prenatal hypoxaemia, and with maternal rubella in first trimester of pregnancy.

Basic physical findings in PDA :

Pulse - High-volume, bounding or water-hammer in character; wide pulse pressure.

Apex beat - Hyperdynamic.

Thrill - Systolic or systolo-diastolic thrill may be palpable at the left 2nd intercostal space.

Heart sounds - S₁ and S₂ are loud, and S₃ may be audible at the apex.

Murmurs - The typical continuous 'machinery' murmur ('Train in tunnel' murmur, Gibson's murmur) with late systolic accentuation is best audible in left infraclavicular area and left 2nd intercostal space. A mid-diastolic apical rumbling murmur may be audible due to increased flow across the mitral valve.

* **Chest X-ray** reveals LVH, LAH, big pulmonary artery, calcification of ductus (in elderly), and rarely huge aneurismal dilatation of ductus (due to infection in ductus).

** ECG shows LVH and LAH; RVH in pulmonary hypertension.

*** **Management** - Premature infants with PDA are treated medically with indomethacin or ibuprofen (prostaglandin inhibitor) in the first week of life to stimulate ductal closure. Surgery is done by ligation (< 5 years), and ligation as well as excision of the patent duct (> 5 years).

Associated anomalies with PDA :

- | | |
|--|------------------------|
| 1. Coarctation of aorta. | 4. Tricuspid atresia. |
| 2. Congenital aortic stenosis. | 5. Pulmonary stenosis. |
| 3. TAPVC and TOF may be associated with. | 6. VSD. |

PDA is lifesaving in :

1. Aortic atresia
2. Pulmonary atresia
3. Severe coarctation of aorta
4. Incomplete aortic arch syndrome
5. Hypoplastic left heart syndrome.

Eisenmenger's syndrome :

Pulmonary hypertension with reversal of shunt (right-to-left) from an initial left-to-right shunt is known as Eisenmenger's syndrome and the causes are:

1. VSD (Eisenmenger's complex) - occurs earlier in life.
2. pda - develops a little later than VSD.
3. ASD - develops late in adult life.

Mechanism: Persistently elevated pulmonary flow (left-to-right shunt) raised pulmonary resistance -> pulmonary hypertension -> progressive, obliterative and irreversible changes occur in the pulmonary vasculature persistent pulmonary hypertension -> reversed right-to-left shunting of blood.

The **symptoms** are breathlessness, fatigue, syncope, angina, haemoptysis or features of CCF.

The **developing signs** are:

1. Central cyanosis (not corrected even by giving 100% oxygen).
2. Polycythemia.
3. Clubbing.
4. Prominent a-wave in neck veins.

5. Features of RVH (outward apex, left parasternal heave and epigastric pulsation) or RVF.
6. Loud and palpable P_2 .
7. Ejection click and ejection systolic murmur due to pulmonary hypertension.
8. Occasionally pansystolic murmur of tricuspid incompetence may be audible.
9. Original murmur (VSD, PDA, ASD) decreases in intensity, duration and may disappear.

Conditions provoking systemic vasodilatation like pyrexia, exercise, hot climate may trigger the right-to-left shunt and worsen the systemic desaturation. Pregnancy and anaesthesia are tolerated very badly by these patients. Haemoptysis is a bad sign in Eisenmenger's syndrome though not very common. The **chest X-ray** is characteristic which reveals enlarged central pulmonary arteries and peripheral 'pruning' of pulmonary vessels (i.e., peripheral $\frac{1}{3}$ rd of lungs have less arterial markings). The ECG shows features of RVH. Heart-lung transplantation remains the only curative treatment.

* PDA with reversal of shunt produces differential cyanosis (feet blue and hands red).

** Remember, Right-to-left shunt gives rise to cyanosis, left-to-right shunt does not.

N.B. : Cyanotic congenital heart diseases are usually given as short and spot cases.

Case 29

FIBROSIS OR COLLAPSE OF THE LUNG

Clinical presentation :

- | | |
|---|---------------------------------------|
| 1. Respiratory distress. | Symptomatology basically depends on : |
| 2. Dry cough. | • Rapidity of development. |
| 3. Pain in the affected chest. | • Amount of lung parenchyma involved. |
| 4. Pyrexia (may or may not be present). | • Presence or absence of infection. |

Describe the outline of findings in this case :

(A) GENERAL SURVEY—

1. Dyspnoea, orthopnoea (if a major bronchus is suddenly obstructed).
2. Central cyanosis.
3. Clubbing Fibrosing alveolitis or collapse developing from bronchogenic carcinoma.
4. Tachypnoea.
5. Tachycardia.

(B) INSPECTION—

1. Flattening of the chest with supraclavicular hollowing, infraclavicular flattening and supra-scapular wasting.
2. **Crowding of ribs and narrowing of intercostal spaces**—Specially in long standing cases.
3. Restricted movement on affected side.
4. Drooping of the shoulder in case of involvement of apex (seen from back).
5. Shifting of apical impulse towards the affected side.
6. Kyphoscoliosis resulting from extensive fibrosis.

(C) PALPATION—

1. **Shifting of trachea and apex beat towards the affected side.**
2. Movement of the chest Diminished movement and reduced expansion on the diseased side.
3. Vocal fremitus—
 - (i) Fibrosis—Diminished.
 - (ii) Collapse Diminished or absent if the bronchus is obstructed, and increased if the bronchus is patent.

(D) PERCUSSION—Impaired resonance or dull note.

(E) AUSCULTATION—

- a) Collapse with obstructed bronchus or fibrosis—Diminished vesicular (fibrosis) or absent breath sound (collapse), diminished or absent vocal resonance, fine crepitations and occasionally rhonchi are heard in fibrosis; rhonchi and crepitations are absent in collapse with obstructed bronchus.

- b) Collapse with patent bronchus—Tubular breath sound, bronchophony with whispering pectoriloquy, and occasionally coarse crepitations are heard.

* *Inflibrosis, the onset is chronic and the chest wall is retracted in comparison to collapse of the lung where the onset is acute and the chest wall is flattened. Drooping of the shoulder, flattening of chest or crowding of ribs (on affected side) are commonly found in fibrosis, and may be seen in collapse of prolonged duration.*

** Past history—Tuberculosis, mumps/measles/whooping cough in childhood, joint pain, symptoms regarding bronchogenic carcinoma should be enquired into.

Types of fibrosis of the lung :

a) *Focal fibrosis*—Coal-worker's pneumoconiosis, asbestosis, silicosis.

b) *Replacement fibrosis*—Tuberculosis, bronchiectasis, radiation fibrosis, lung abscess, pulmonary infarction, necrotizing pneumonia.

c) *Interstitial fibrosis*—This is the fibrosis of the alveolar walls, which affects both lungs diffusely (fibrosing alveolitis). It is commonly associated with systemic collagen diseases like rheumatoid arthritis, scleroderma etc.

* Patient is more symptomatic in a) and c) but physical signs are more pronounced in b).

** Fibrosis of the lung of cardiac origin are mitral stenosis and multiple pulmonary infarcts.

What is 'diffuse fibrosis' of the lung ?

1. Acute—Hamman-Rich disease (acute onset, progressive course, unknown aetiology).
2. Chronic—Fibrosing alveolitis.

Diffuse fibrosis of the lung (chronic) is diagnosed clinically but confirmed by lung biopsy. The findings are bilateral and diffuse (not localised).

Classical features of fibrosing alveolitis at the bedside :

1. Progressive disabling dyspnoea, orthopnoea; tachypnoea; persistent dry cough; weight loss and fatigue.
2. Constitutional symptoms like arthralgia (rare).
3. Central cyanosis, polycythemia.
4. Clubbing.
5. Bilateral diminution of chest movement; diminished vesicular breath sound.
6. Leathery crepitations (velcro crepitations linked to unzipping of velcro)—Chiefly end-inspiratory crepitations, uninfluenced by cough, heard particularly over both the lower zones at back (bi-basal crepitations).
7. Signs of pulmonary hypertension, RVF or chronic cor pulmonale in late stages.

* These patients are usually corticosteroid-responsive.

How do you classify collapse of the lung ?

(A) *ABSORPTION OR ACTIVE COLLAPSE*—The bronchus is either obstructed by mucus plugs or bronchial casts, foreign body, blood, neoplasm (endobronchial), or compressed from outside by enlarged glands, aneurysm, neoplasm (exobronchial), or by neoplasm arising within the bronchial wall (intra-bronchial). The trachea is shifted to the same side due to more negative intrapleural pressure.

(B) *COMPRESSION OR PASSIVE COLLAPSE*—The bronchus remains patent, and is developed as a result of pleural effusion, empyema, pneumothorax, hydropneumothorax, pyopneumothorax or very large neoplasm. The trachea is pushed to the opposite side as a result of positive intrapleural pressure.

* Previously absorption collapse was known as atelectasis of lung.

** In clinical practice, unless or otherwise qualified, collapse means absorption collapse (i.e., the model case described above). Whereas, signs of compression collapse literally means signs of pleural effusion/pneumothorax/hydropneumothorax where the underlying lung remains silent.

*** Collapse may again be divided by ;

- Central (mass lesion)—bronchus is obstructed: all signs of collapse are dampened.
- * Peripheral—bronchus remains patent; bronchial breath sound and bronchophony +.

Common causes of absorption collapse in relation to age :

- a) Neonates—meconium aspiration,
- b) Children—foreign body aspiration,
- c) Adults—bronchial adenoma,
- d) Old age—bronchogenic carcinoma.

What is 'middle lobe syndrome'?

It is the recurrent or persistent atelectasis of right middle lobe caused by compression of bronchus as a result of enlarged lymph nodes arising out of tuberculosis or malignancy. The diagnosis is confirmed by CT scan or bronchoscopy.

Clinical features of bronchiectasis :

1. Symptoms—
 - a) Cough with expectoration—Copious, purulent, foetid, often related to change of posture, most prominent in the early hours of the morning while arising from bed or at the time of retiring to bed.
 - b) Haemoptysis.
 - c) Systemic symptoms like febrile episodes, malaise, anorexia, loss of weight.
 - d) Breathlessness.
 - e) Pain in the chest (due to pneumonia or dry pleurisy).
2. General survey—
 - a) Dyspnoea.
 - b) Clubbing.
 - c) Anaemia, fever, halitosis.
 - d) Central cyanosis in bilateral and extensive disease.
 - e) Undernutrition.
3. Examination of the chest—There may be no abnormal sign in the chest. The signs depend on the size of affected bronchi, airway patency and viscosity of secretions. One may get the signs of (i) bronchitis (ii) fibrosis (iii) consolidation (iv) collapse (v) cavitation or, rarely (vi) pleural effusion.

The physical signs often **change after a large bout of cough**. Breath sound may be vesicular with prolonged expiration to high-pitched bronchial (tubular) in type. Sharp metallic or bubbling 'leathery', coarse crepitations are characteristic. All the **signs are predominantly basal and bilateral**.

Clinical features of pulmonary tuberculosis (post-primary) :**(A) Symptoms :**

- a) Evening rise of temperature, anorexia, loss of weight, lassitude, night sweats, palpitation.
- b) Persistent cough; sometimes there is sputum production (in advanced stage).
- c) Haemoptysis.
- d) Chest pain (due to pleurisy).
- e) Breathlessness.

(B) Signs :

Initially there may be no abnormal physical sign in the chest. The earliest sign is often post-tussive crepitations heard in the apex of the lung. Gradually the physical signs of tuberculous consolidation, cavitation, fibrosis, pleurisy, pleural effusion or spontaneous pneumothorax may develop.

Clinical stigmata of evidence of past or present tuberculous infection :

- Erythema nodosum.
- Phlyctenular keratoconjunctivitis.
- Old scar or sinus reflecting the foot prints of tuberculous lymphadenopathy.
- Scrofuloderma.
- Beaded and thickened spermatic cord.
- Gibbus

Effects of asbestos exposure in respiratory system :

At least 10 years of moderate to severe exposure to asbestos has occurred before the disease becomes overt. It affects persons working in asbestos mines, textile industries, manufacturer of fireproof and shipyard workers. The manifestations are :

1. Asbestosis (progressive pulmonary fibrosis)—Dry cough, chest tightness, dyspnoea on exertion, clubbing and end-inspiratory crepitations at lung bases.
2. Benign pleural plaques, diffuse pleural fibrosis.
3. Benign pleural effusion.
4. Bronchogenic carcinoma—Usually adenocarcinoma.
5. Mesothelioma of pleura (and peritoneum)—Produces malignant pleural effusion.
6. Laryngeal carcinoma.

* So, proper attention must be paid while enquiring for occupational history, both present and past.

** One year's exposure to asbestos in teens may even result in mesothelioma in the fifth decade.

What is 'pack year' ?

It is the duration of smoking in years multiplied by number of packets of cigarettes smoked per day, e.g., one pack of cigarette (containing 20 cigarettes) smoked per day for 30 years make 30 pack years. The importance lies in the fact that risk for development of bronchogenic carcinoma is much increased when the pack years exceed 40. Pack year has also direct relationship with development of COPD.

Position of mediastinum in collapse of the lung :

In a wider sense, collapse of the lung means absorption collapse.

(A) Absorption collapse—Mediastinum is pulled to the affected side.

(B) Compression collapse—Mediastinum is pushed to the opposite side.

* Read the section on 'Pleural effusion' for better understanding.

Bronchopulmonary segments in the lung :

Bronchopulmonary segment is a part of lung tissue supplied by one segmental bronchus, along with corresponding pulmonary artery and pulmonary vein. There are 3 lobes (upper, middle and lower) in right, and 2 lobes (upper and lower) in left side of lung. Right lung have 10 and left lung have 9 bronchopulmonary segments (medial basal segment is usually missing in left side).

Investigations you like to perform (fibrosis or collapse) :

1. Blood—Raised ESR, polycythemia; rheumatoid factor, antinuclear antibodies or circulating immune complexes may be found in fibrosing alveolitis.
2. Chest X-ray (PA view; lateral view is also required to determine the segment or lobe involved)—
 - (i) Homogeneous shadow of the collapsed lung.
 - (ii) Displacement of mediastinum towards the affected side.
 - (iii) Retraction of hemithorax on the affected side.
 - (iv) 'Honeycomb lung' in fibrosing alveolitis,
 - (v) Tenting of diaphragm in basal collapse or fibrosis in the base.
 - (vi) In compression collapse—The aetiology of compression is seen in the X-ray plate e.g., pneumothorax, pleural effusion.
3. High resolution CT scan—Shows honeycombing and scarring, which are marked peripherally in both the lungs. CT scan helps in early and confirmed diagnosis.
4. Lung function tests (specially in fibrosis)—Shows restrictive type of defect i.e., FEV/FVC is approximately 90% (in obstructive defect i.e., in bronchial asthma, FEV/FVC is near about 40-50%).
5. Bronchoscopy—To identify the cause of obstruction or to get a biopsy.
6. Rarely lung biopsy is needed to diagnose fibrosing alveolitis (i.e., the biopsy specimen shows patchy foci of interstitial fibrosis).

NB In a 'Short case', a student has to examine the patient for **general survey and one particular system** asked for: the candidate is usually not allowed to take the history and thus, need not write the case history sheet. Commonly, the time allotted is 10-15 minutes.

Case 30

SUPERIOR MEDIASTINAL SYNDROME

Synonyms :

1. Superior vena caval (SVC) syndrome.
2. Superior vena caval obstruction.
3. Reversed congestive cardiac failure (i.e., as upper half of the body is swollen).

What is SVC syndrome ?

There is obstruction of SVC due to compression and/or infiltration by superior mediastinal tumours which result in characteristic features like headache, breathlessness, plethora with oedema of the face, dilatation of collateral veins in upper thorax and conjunctival oedema.

Common causes of SVC syndrome :

Malignant diseases are the principal cause of this syndrome and therefore, the prognosis is very bad. The causes are,

1. Bronchogenic carcinoma (75%).
2. Lymphoma (20%).
3. Giant aortic aneurysm.
4. Retrosternal goitre.
5. Tumours of the thymus.
6. Fibrosing mediastinitis (methysergide-induced).
7. Metastatic tumour e.g., from carcinoma of breast.
8. Dermoid cyst.
9. Persistent left-sided SVC.
10. Thrombosis of SVC.
11. Teratoma.
12. Pericardial cyst.

Different modes of presentation (symptoms) :

The mediastinal mass usually compresses three tubes, three nerves and three vessels. In bronchogenic carcinoma and lymphoma, the mediastinal structures are involved by spread to mediastinal lymph nodes and sometimes by direct extension of the tumour itself.

(A) TUBES :

- a) Oesophagus—The patient may present with dysphagia and as it is a mechanical compression of the oesophagus, patient complains of difficulty in swallowing food mainly with solids (later on solid plus liquid).
- b) Trachea Respiratory distress, stridor and paroxysmal cough.
- c) Bronchus—As such, obstruction of the bronchus from a mediastinal mass (due to bronchogenic carcinoma) is not very common. *The manifestations of obstruction depend on the nature of obstruction (complete or partial), absence or presence of secondary infection, and its effect on pulmonary Junction.* When a large bronchus is 'completely' obstructed, there is absorption collapse or atelectasis, and the patient may complain of breathlessness and pain chest. If the collapse involves a smaller portion of lung, there may be no respiratory difficulty. In case of 'partial' obstruction of bronchus, there is development of obstructive emphysema (the airflow resistance is increased in expiration than during inspiration and causes overdistension of the lung, lobe or segment) and thus, respiratory embarrassment may be present. Secondary infection of the lung is very common in this situation and the patient may present with features of pneumonia

(B) NERVES:

- a) Recurrent laryngeal nerve The left-sided nerve is commonly involved due to its lower station than the right. The right-sided recurrent laryngeal nerve remains high up and is usually not compressed by the mediastinal mass. The patient presents with,
 - (i) Hoarseness of voice,
 - (ii) Bovine cough (explosive nature of cough is lost), and
 - (iii) Stridor (common in bilateral palsy).
- b) Phrenic nerve—There may be breathlessness (mainly in supine position) and sometimes, respiratory failure is present due to diaphragmatic paralysis.

U Cervical sympathetic trunk (Homer's syndrome)—Patient complains of partial drooping of the upper eyelid and loss of sweating on the affected side of face

(C) VESSELS :

- a) Superior vena cava—the patient usually complains of,
 - (i) Swollen and puffy face,
 - (ii) Headache,
 - (iii) Visual disturbances,
 - (iv) Alteration in the state of consciousness, black-out, and
 - (iv) Rarely convulsions.
- b) Azygos vein—Symptoms of pleural effusion (on the side of compression, i.e., right-sided).
- c) Lymphatics—Thoracic duct or small lymph channels may be obstructed. There are presence of,
 - (i) Parietal oedema of upper thorax, and
 - (ii) Symptoms of pleural effusion (chylothorax).

* Pericardial effusion may develop.

Physical findings in a case of SVC syndrome (signs) :**(A) TUBES :**

- a) Oesophagus—Dysphagia may be demonstrated if the patient is allowed to eat some biscuits.

- b) Trachea—There may be intercostal suction, central cyanosis and presence of stridor.
- c) Bronchus—
 - (i) Signs of absorption collapse (in obstructed bronchus) is found, i.e., there is diminished movement on the affected chest, shifting of the mediastinum to the affected side, impaired note on percussion, absent breath sound with diminished or absent vocal resonance, and usually without any adventitious sound.
 - (ii) Rarely, the cause of SVC syndrome (e.g., bronchogenic carcinoma or lymphoma) may produce pleural effusion and thus, there may be compression collapse. The signs are diminished movement on the affected chest, shifting of the mediastinum to the opposite side, stony dullness on percussion, diminished vesicular or tubular breath sound, diminished vocal resonance or bronchophony, aegophony, whispering pectoriloquy with few coarse crepitations (usually the mediastinum remains in the middle or on the same side, and rarely on the opposite side — for better understanding, read the section on Pleural effusion).
 - (iii) Rarely, a large mediastinal lymph node (at bifurcation of trachea) in lymphoma or the mass in bronchogenic carcinoma may transmit the tracheal sound and as a result of this, there is production of tubular (bronchial) breath sound, whispering pectoriloquy and rarely aegophony (heard in the interscapular region). This is known as D'Espine's sign (posterior).

* Dull note on percussion over 2nd intercostal space anteriorly (right or left) with presence of tubular (bronchial) breath sound is anterior D'Espine's sign.

(B) NERVES :

- a) Recurrent laryngeal nerve—
 - (i) Ask the patient his name or address, and observe the hoarseness of voice.
 - (ii) Ask the patient to cough for demonstration of bovine cough.
 - (iii) Indirect laryngoscopy shows the position of the affected vocal cord (commonly left-sided) in paramedian position (i.e., cadaveric position) during phonation.
- b) Phrenic nerve—Read the section on 'Hydropneumothorax' for phrenic nerve palsy.
- c) Cervical sympathetic trunk—Horner's syndrome is characterised by,
 - (i) Pseudoptosis,
 - (ii) Constriction of pupil (miosis),
 - (iii) Anhidrosis (lack of sweating) of ipsilateral face, neck, arm and upper chest,
 - (iv) Enophthalmos, and
 - (v) Loss of ciliospinal reflex.

(C) VESSELS :

- a) Superior vena cava—
 - (i) **Dilatation of tortuous collateral veins in upper chest, arm, neck** and often in temple region of the face; even small venules may be prominent over chest wall.
 - (ii) Oedema of the face (moon face), neck, arm and upper thorax; engorged sublingual veins.
 - (iii) Plethora of the face (often there is cyanotic hue of face, neck, arm and hands).
 - (iv) **Engorged and non-pulsatile internal jugular vein (with negative hepato-jugular reflux).**
 - (v) Conjunctival oedema (chemosis), and subconjunctival haemorrhage.
 - (vi) The flow of blood in the collateral veins is from 'above downwards' (read the section on 'Prominent veins with venous hum' for demonstration of venous flow).
 - (vii) There may be disturbed sensorium.
 - b) Azygos vein—Rarely, there may be pleural effusion (on the side of compression of the azygos vein, i.e., right-sided).
- c) Lymphatics—
 - (i) Chylothorax.
 - (ii) Non-pitting oedema of upper thorax (front and back).

* Rarely, there is presence of pericardial effusion in SVC syndrome. On an anatomical basis, right-sided tumours produce this syndrome much more frequently.

** Two other features of SVC syndrome are bilateral papilloedema and exophthalmos. Remember, all the features in this syndrome increase after leaning forward (rel : breathlessness in pericardial effusion often diminishes after adopting Mahammedan's prayer position).

*** Chest wall veins are not prominent if SVC obstruction occurs above the joining of azygos vein.

**** Q_{ne} must look for cervical lymphadenopathy, clubbing, radiation mark in chest and pulsus paradoxus.

Differential diagnosis you will consider :

Mention the causes of 'Moon face'. Other D/D of SVC syndrome are constrictive pericarditis, CCF and chronic cor pulmonale.

How do you like to investigate the patient ?

1. Blood examination—Hb, TC, DC, ESR, sugar (F).
2. Chest X-ray—PA view (lateral view helps to identify the anatomical site of the tumour).
 - (i) **Mediastinal widening.**
 - (ii) Atelectasis.
 - (iii) Dense irregular hilar opacity which is extending peripherally (bronchogenic carcinoma).
 - (iv) Pulmonary parenchymal opacity (bronchogenic carcinoma).
 - (v) Pleural effusion, elevation of the hemidiaphragm (phrenic nerve palsy), rib erosion.
 - (vi) Rarely, pericardial effusion.
3. Indirect laryngoscopy to see the paramedian position of left-sided vocal cord on phonation in a patient with hoarseness of voice.
4. Cytological examination of sputum, bronchial brushings or washings for malignant cells.
5. FNAC or excision biopsy of scalene node, Virchow's gland or any palpable lymph node.
6. Pleural fluid aspiration and pleural biopsy.
7. Scanning of lung or thyroid (by using I¹³¹ or technetium 99m for retrosternal goitre), or **CT scan of the thorax with FNAC**, if possible.
8. Barium swallow examination shows oesophageal displacement or compression.
9. Bronchoscopy, oesophagoscopy and mediastinoscopy are usually contraindicated in the acute stage due to risk of bleeding. If the obstruction is relieved by treatment (radiotherapy or corticosteroid), one skilled clinician may go through, the investigations for tissue diagnosis.

How you are going to manage a patient of SVC syndrome ?

The SVC syndrome is a serious medical problem and should be managed promptly. Unless the clinical examination and non-invasive diagnostic procedures suggest a benign cause of obstruction, or unless there is availability of tissues elsewhere in the body for biopsy (e.g., lymph node biopsy), the patient should receive irradiation or chemotherapy (cyclophosphamide) before attempts are made to come to a tissue diagnosis. Elevation of head and oxygen therapy may give temporary symptomatic relief; ventilatory support may be necessary. Corticosteroids and diuretics are often used initially to reduce oedema at the site of obstruction. Tracheal obstruction needs emergency radiotherapy. When a tissue diagnosis is made afterwards, the patient is managed accordingly.

What is inferior vena caval (IVC) syndrome ?

- a) Extraluminal compression—Ascites, tumours in the abdomen and pelvis, tuberculous plastic peritonitis, pregnancy.
- b) Intraluminal compression — Malignancy of kidney or adrenals, thrombosis extending from pelvic veins, membranous obstruction of IVC, OC pills, myeloproliferative diseases, hypercoagulable states.

Clinical features—

1. Pitting oedema of legs with dilated varicose veins; local cyanotic hue.
2. Collateral veins are seen over the abdomen, flanks and back.
3. Swollen and oedematous buttocks and groin.
4. Sometimes there is presence of ascites (ascites follows pedal oedema).
5. Direction of blood flow is from 'below upwards'.
6. Stasis ulcer on the legs with pigmentation in chronic cases.

Conclusion :

In a patient of SVC syndrome, obstruction of bronchus is rarely seen in comparison to compression of vein and nerve (very common). In the SVC syndrome, both direct (dull note on sternal percussion and D Espine's sign) and indirect (vein and nerve compression) effects are observed though the indirect effects are more commonly encountered. Engorged and non-pulsatile neck vein is a very important clinical clue. Always percuss the sternum in a suspected case of SVC syndrome.

A typical patient may have dyspnoea, stridor, puffy and plethoric face with periorbital oedema, and tortuous veins over chest wall.

Case 31

ASCITES

What is ascites ?

Ascites refers to collection of 'free' fluid within the peritoneal cavity. Normally, there is virtually no fluid present in the peritoneal cavity. The Greek word 'askitos' means bag or sac.

Causes of ascites :

(A) ASCITES WITH ANASARCA—

1. Congestive cardiac failure (CCF).
2. Nephrotic syndrome.
3. Hypoproteinaemia with severe anaemia (nutritional).
4. Pericardial effusion.
5. Constrictive pericarditis.
6. Myxoedema.
7. Filariasis.
8. Protein-losing enteropathy (due to hypoproteinaemia).
9. Epidemic dropsy.

(B) ASCITES WITHOUT ANASARCA (ONLY ASCITES)—

1. Cirrhosis of liver (may lead to anasarca).
2. Peritonitis—
 - (i) Acute pyogenic peritonitis.
 - (ii) Tuberculous peritonitis.
 - (iii) Malignant peritonitis.
 - (iv) Spontaneous bacterial peritonitis (SBP).
3. Portal vein thrombosis.
4. Meig's syndrome.
5. Pancreatic ascites (e.g., from acute pancreatitis).
6. Hepatic vein thrombosis (Budd-Chiari syndrome).
7. Rupture of hollow viscus within abdomen.
8. Lymphoma or leukaemias.
9. Chylous ascites.
10. Haemoperitoneum following trauma.
- IX. Rare - Vasculitis, peritoneal dialysis, IVC obstruction.

* Common causes of ascites are 1, 2, 3 of (A) and 1, 2 of (B).

** In hypoproteinaemia with severe anaemia, oedema is out of proportion to ascites.

*** Ascites in cirrhosis of liver is mainly due to hepato-cellular failure (i.e., hypoalbuminaemia) and partly due to portal hypertension.

Points to be asked in history in a patient with ascites :

1. Onset (sudden or gradual) and progress (slowly growing, stationary or progressive).
2. Past history of jaundice, haematemesis or melaena, alcoholism, fever (see below).
3. Pain and tenderness (tuberculous peritonitis, acute pancreatitis).
4. Loss of appetite.
5. Loss of weight.
6. Cough with or without haemoptysis.
7. Swelling of legs or face.
8. Respiratory distress (huge ascites, CCF, pericardial diseases).
9. Prolonged diarrhoea (protein-losing enteropathy), alteration of bowel habit (large gut malignancy) or alternate diarrhoea and constipation (intestinal tuberculosis).
10. Any sort of oliguria, paraesthesia in lateral thigh, or heart burn present or not.

Important past history in a patient with ascites :

- | | |
|-------------------------------|--|
| 1. Alcoholism. | 6. Haematochezia. |
| 2. Jaundice. | 7. Pain abdomen (acute pancreatitis). |
| 3. Haematemesis or melaena. | 8. Loss of weight (malignancy). |
| 4. Alteration in bowel habit. | 9. Past H/O rheumatic fever or respiratory distress. |
| 5. Tuberculosis. | 10. Fever (tuberculous or pyogenic peritonitis). |

Why do you say ascites in this case ?

1. There is generalised swelling of abdomen; particularly the flanks are full; umbilicus is everted (or flushed with the abdominal wall) with a transverse slit. Maximum girth of abdomen is at the level of umbilicus.
2. Fluid thrill is present.
3. Shifting dullness confirms the diagnosis of ascites.

What are the clinical features of ascites ?**Symptoms—**

1. Progressive swelling of abdomen with gain in weight.
2. Aching pain all over the abdomen due to stretching; bloated feeling in abdomen.
3. Dyspnoea and even orthopnoea (in massive and rapid collection of fluid).
4. Swelling of lower limbs (due to CCF, cirrhosis of liver or functional IVC obstruction).
5. Paraesthesia in the distribution of lateral femoral cutaneous nerve (meralgia paraesthetica).
6. Oliguria.
7. Dyspepsia, heart burn (due to GERD).

Signs —**(A) GENERAL SURVEY—**

1. The patient may be in propped-up position due to dyspnoea.
2. Obvious swelling of abdomen.
3. Neck veins may be engorged (due to hypervolaemia).
4. Bipedal pitting oedema.

(B) ABDOMEN—

1. Swelling of abdomen and there is fullness in the flanks. The skin looks stretched and shiny.
2. Divarication of recti (if the patient tries to get up from supine position against resistance, the linea alba bulges between the two recti) may be present; there may be presence of abdominal (umbilical) herniation or abdominal striae (striae albicantes). Subcostal angle is wide, and lower ribs are pushed upward and outward. There is prominence of hypogastrium in erect posture.
3. There may be prominent veins around umbilicus or in epigastrium where the blood flow is away from the umbilicus (portal hypertension—in case of cirrhosis of liver)—more common. Prominent veins in flanks of abdomen may also be seen where the blood flow is from below upwards (functional IVC obstruction by ascites itself—less common).
4. Umbilicus—Flushed or everted with transverse slit; may have umbilical hernia.
5. Fluid thrill—Present.
6. Shifting dullness—Present.
7. Measurements—
 - a) Maximum girth of abdomen is at the level of umbilicus.
 - b) Spino-umbilical distance (line joining anterior superior iliac spine and umbilicus) is more or less equal on both sides.
 - c) The distance between xiphisternum and umbilicus is more than the distance between umbilicus and symphysis pubis.
8. Auscultation—Venous hum over the distended veins around umbilicus may be audible in cirrhosis with portal hypertension.

;C) RESPIRATORY SYSTEM—

1. Basal collapse—Diminished movement, impaired note on percussion, diminished vesicular breath sound and crepitations may be heard in the lower part of chest on both sides.
2. Hydrothorax (commonly right-sided) due to leakage of ascitic fluid through defects (pores) in the right dome of diaphragm for lymphatic channels; may be bilateral.

(D) CVS—

1. Apex beat—Deviated upward and outward.
2. There may be diffuse pulsation over the precordium.
3. Soft systolic murmur in pulmonary area may be present (due to associated severe anaemia).

(E) NERVOUS SYSTEM—

Nothing particular. There may be features of hepatic encephalopathy.

(F) GENITOURINARY SYSTEM—Scrotal oedema or hydrocele may be evident as secondary effects of ascites (rare) or as a clue to aetiological (e.g., nephrotic syndrome) diagnosis of ascites.

N.B. : With the above mentioned signs (for ascites per se), there are presence of other features due to

specific aetiology of ascites. Thus, one should always be careful in searching the other stigmata of cirrhosis of liver, CCF etc in a case of ascites. P/R and P/V examinations should be done.

* A **long-standing ascites** is recognised by striae albicantes, divarication of recti (may be associated with visible peristalsis underneath), umbilical hernia and puncture marks.

** Palpate liver, spleen or any other abdominal viscera by dipping method, in the presence of ascites.

Examination for fluid thrill :

1. The patient lies supine with thighs flexed. Either the patient or a third person (in the case of a child patient) will *put his ulnar border of right hand vertically, with fingers directing caudally, over the abdomen in the midline* (sagittal plane) firmly to prevent transmission of impulse or vibration through the abdominal parietis. The impulse passing through the posterior abdominal wall is dampened by the bed.
 2. Place the left palm over the left flank and sharply tap or flick the right flank with the fingers of right hand; a fluid thrill or 'shock wave' is felt by the left palm as a definite impulse. The same manoeuvre is now done by tapping the left flank while the impulse is felt over the right flank.
- * Preferably, the bladder should be evacuated before performing the test.

Where do you get fluid thrill ?

Fluid thrill is present in the presence of fluid in the peritoneal cavity **whether it is encysted or free**. Thus, fluid thrill may be found in,

- | | |
|---------------------|------------------------------------|
| 1. Ascites (tense), | 4. Rarely in large hydronephrosis, |
| 2. Ovarian cyst, | 5. Any large intra-abdominal cyst, |
| 3. Hydramnios, | 6. Obesity (false positive). |

Essential criteria for positive fluid thrill :

1. Fluid should remain **under tension** (most important).
 2. Large amount of fluid (it is often said that at least 2 litres fluid is necessary to elicit fluid thrill).
- N.B. ; Fluid thrill may not be present if there is small collection or the fluid is not under tension.

Is fluid thrill a definite sign for ascites ?

No.

Surest sign for presence of free fluid in the peritoneal cavity (ascites) :

'Shifting dullness' (in addition, paracentesis abdominis also confirms the diagnosis).

How do you elicit shifting dullness ?

Principle ; The presence of ascites can be confirmed by altering the posture of the patient and demonstrating a change in the position of air-fluid interface (i.e., the distinct transition zone between tympany and dullness). Shifting dullness has both good sensitivity and specificity.

This method may also be called as 'shifting tympanicity'.

1. The patient lies supine with thighs flexed. The patient is asked to evacuate his/her bladder (full bladder will unnecessarily hamper the midline percussion). Try to exclude pregnancy.
2. Now palpate the abdomen (by dipping method) for any visceromegaly (liver, spleen, kidney lump etc.). If any viscus is enlarged, try to avoid percussion over them.
3. Starting from the epigastrium, percuss (usually light to medium percussion) in the midline from above downwards. Note the maximum point of tympanicity. Usually it is somewhere around the umbilicus.
4. Now percuss laterally to one side (suppose, the right side) from the maximum point of tympanicity noted in the midline, keeping the pleximeter finger parallel to long axis of abdomen or arbitrary border of the fluid level. When you get a dull note, mark it with a skin pencil but keep on percussing towards the flanks to diagnose that the dullness is continuous and not a localised one due to colonic growth, faecolith etc., and again return to the first noted point of dullness.
5. Now turn the patient to left lateral position, keeping the fingers in the point of dullness noted in the right flank. Turning is done in such a way that your fingers in the right flank become the highest point of the patient's body. Now wait for few seconds (at least 10 seconds, usually 30 seconds) for the intestine to float up.
6. Percuss the dull point noted in the right flank, which will reveal tympanitic resonance. Go on percussing upto the end point of the right flank which also shows tympanitic resonance; next go upto the midline and now the midline which was tympanitic initially will be dull on percussion.

So, the dullness in the right flank changes to tympanitic note and the midline becomes dull from the original tympanicity. This is shifting dullness.

7. The same procedure is repeated in the left flank starting from the midline.

Minimal fluid required for demonstration of shifting dullness :

It is said that at least 1 / 2 to 1 litre of fluid is required to demonstrate shifting dullness.

Absent shifting dullness in the presence of fluid in the abdomen :

The possibilities are :

1. Fluid is encysted i.e., ovarian cyst or loculated ascites in tuberculous peritonitis.
2. Small collection of free fluid.
3. Rarely in massive tense ascites.

Massive splenomegaly or hepatomegaly with ascites : how to elicit fluid thrill ?

It is often very difficult to form an opinion regarding fluid thrill in the presence of visceromegaly. One should not place the palm over the enlarged organ (may dampen the 'shock wave' of positive fluid thrill), rather one should tap from that side while the palpating hand is kept on the other side (patient's hand remains vertically in the midline). In gross hepatosplenomegaly, fluid thrill may remain inconclusive.

Shifting dullness present but there is no free fluid in the abdomen :

False positively in paralytic ileus (rare).

What is meant by free fluid' in ascites :

The fluid shifts or changes its position with the intestinal air with change of posture which never occurs in encysted fluid and thus, shifting dullness is never found in encysted fluid in the abdomen.

In performing shifting dullness why do we percuss the midline first ?

While the patient lies supine, the intestine floats in the midline (in health as well as in ascites), which gives tympanitic note on percussion. To follow the cardinal rules of percussion, we percuss the midline first (tympanitic) and then the flanks (dull) —i.e., we percuss from 'more resonant to less resonant' area.

Result of percussion over an ovarian cyst :

Fluid thrill may be present but shifting dullness is always absent. In a large ovarian cyst, the midline will be totally dull on percussion. But in a moderate ovarian cyst, upper part of the midline may be tympanitic though the shifting dullness is always absent. In an ovarian cyst, flanks are resonant on percussion.

* If the midline is totally dull on percussion (e.g., large ovarian cyst), it is not necessary to perform the next steps of shifting dullness i.e., if the midline is dull on percussion, shifting dullness will be absent.

** Absence of fluid thrill and/or shifting dullness does not exclude the presence of ascites.

Clinical detection of 'small amount' of free fluid :

(A) **PUDDLE SIGN**—First percuss the abdomen in supine position where you will get a tympanitic note in the midline.—Now place the patient on the hands and knees (knee-elbow position) for 5 minutes and percuss (with difficulty) over the dependent part of abdomen (near umbilicus) which now reveals a dull note due to shifting of fluid. Generally 300-400 ml of fluid may be clinically elicited by puddle sign and even as little as 120 ml of fluid can be detected by this method. This sign is actually elicited by ausculto-percussion (placing the stethoscope over the dependent part and flicking the abdominal wall repeatedly to elicit changes in character of percussion note) which is a very complicated method. Puddle' literally means 'a small pool of muddy water'.

(B) **PER RECTAL EXAMINATION**—Boggy and fluctuant swelling in the dependent part due to presence of free fluid.

(C) **SAME-SIDED SHIFTING DULLNESS**—Very difficult to demonstrate. During the examination by conventional method of shifting dullness, turn the patient to the side of flank percussion and not to the opposite side. Now percuss the flank with difficulty which gives a dull note (here, flanks are originally resonant in supine position due to 'small' collection of free fluid).

Massive hepatosplenomegaly with ascites : how to elicit shifting dullness ?

First percuss the midline in supine position, which will elicit a tympanitic note. Now ask the patient to sit and again percuss the midline from above downwards. In the presence of free fluid in the abdomen, the lower part of midline will be dull on percussion in sitting position (bladder must be evacuated). Actually, in the presence of gross visceromegaly, clinical detection of ascites becomes very difficult.

What is 'horse shoe shaped' dullness in ascites ?

In moderate ascites, the distribution of fluid in supine position of the patient is confined to the flanks, dependent parts and the hypogastrium. Thus, in supine position the epigastrium and umbilical areas are tympanitic on percussion (due to floating of intestine) but a horse-shoe shaped dullness with concave upper border is elicited due to dull flanks and dull hypogastrium. Percuss the abdomen in various directions from the central area of tympanicity (i.e., the umbilical region) to outwards, and demonstrate the horse-shoe shaped dullness.

Unilateral shifting dullness :

This is found in splenic rupture and is known as Ballance's sign. The blood present in the left flank becomes clotted (near the spleen) and does not shift to right side in right lateral position but the blood present in the right side (haemoperitoneum) is shifted to the left side.

Table 20 : Bedside differentiation between ascites and ovarian cyst

Features	Ascites	Ovarian cyst
1. Uniformity of swelling	1. Symmetrical	1. Asymmetrical
2. Any border seen	2. Never	2. Definite upper border is visible
3. Measurements	3.	3.
(i) Maximum girth	(i) Around umbilicus	(i) Usually below the umbilicus
(ii) Spino-umbilical distance	(ii) Equal on both sides	(ii) Not equal
4. Distance between :	4.	4.
a) Xiphisternum to umbilicus, and	a > b	b > a
b) Umbilicus to symphysis pubis		
5. Flanks	5. Full	5. Not full
6. Umbilicus	6. Everted and the slit is transverse	6. Everted and the slit is vertical
7. To 'get above' the swelling	7. Not possible	7. Usually possible
8. Percussion over flanks	8. Dull	8. Tympanitic
9. Shifting dullness	9. Present	9. Absent
10. Per vaginal examination	10. Nothing characteristic	10. Ovarian cyst is diagnosed

* Rarely, it is seen that midline percussion evokes dull note in a case of very tense ascites where the intestine is compressed (may have short mesentery too) and can not come in front. Remember, the maximum bulge in ascites is transverse in contrast to ovarian cyst where it is antero posterior.

** 'Laughing umbilicus'—When the slit of umbilicus becomes transversely stretched.

*** Ascites may be classified into mild, moderate, or tense according to the amount of fluid collected.

Abdominal measurements in ascites :

1. Abdominal girth at the level of umbilicus (maximum girth is at this level in ascites; in ovarian cyst, it is below umbilicus)—periodic measurement is helpful to assess prognosis in ascites.
2. Spino-umbilical distance—measurement is taken from umbilicus to anterior superior iliac spine on either side. Normally they are equidistant (also in ascites). Unequal distance is characteristic of tumour originating from pelvic organ (e.g., ovarian cyst).
3. Distance between xiphisternum to umbilicus, and umbilicus to symphysis pubis—in health, umbilicus lies in midposition of xiphisternum and symphysis pubis. Umbilicus is displaced down in ascites and upper abdominal mass, and displaced up in ovarian cyst or pelvic tumours.

D/D of bulging or distension of abdomen :

Commonly this is due to "7 F"—

1. Fat—Obesity (inverted umbilicus with absent fluid thrill and shifting dullness).
2. Faeces—Megacolon or low gut obstruction (symmetrical enlargement with visible peristalsis).
3. Foetus—Pregnancy (foetal parts are palpable; central dullness).
4. Flatus—Gaseous distension (flanks are not bulged; tympanitic note all over the abdomen).
5. Fluid—Ovarian cyst, ascites.

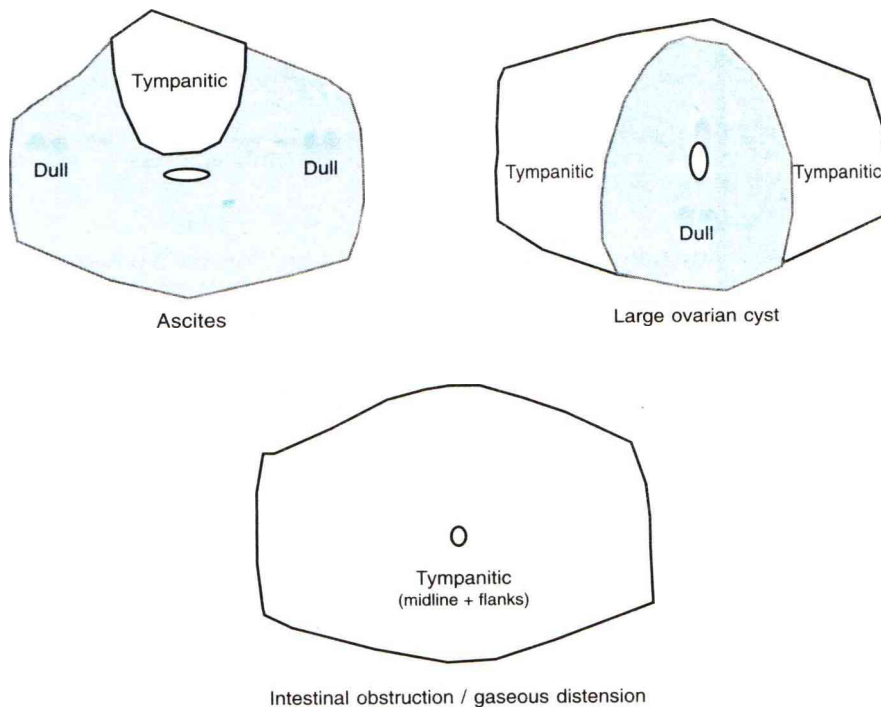


Fig. 1 : Diffuse enlargement of abdomen

6. Full (urinary) bladder (flanks are tympanitic, palpation causes pain or discomfort and desire for micturition; usually rounded cystic swelling in hypogastrium which is dull on percussion; shifting dullness is absent).
7. Fibroid or fatal tumour (e.g., polycystic kidney or massive splenomegaly).

- * Distension of abdomen is of two types :
- a) Generalised (mentioned above), and
 - b) Localised (visceromegaly, neoplasm).

Percussion note in different clinical conditions :

1. Ascites—Midline tympanitic, flanks dull.
2. Ovarian cyst, distended bladder and gravid uterus—Midline dull, flanks tympanitic.
3. Gaseous distension (e.g., intestinal obstruction)—Midline as well as flanks are tympanitic.
4. Obesity—Midline as well as flanks are dull.

Ascites appearing before oedema feet, or ascites > oedema feet (ascites precox):

1. Cirrhosis of liver.
2. Tuberculous or malignant peritonitis.
3. Constrictive pericarditis or restrictive cardiomyopathy.

- * In other conditions (CCF, nephrotic syndrome etc) oedema precedes ascites.

Describe the normal position of umbilicus :

1. Umbilicus lies more or less in the midway between xiphisternum and symphysis pubis.
2. Normally it is inverted and slightly retracted.
3. Normal umbilicus is a puckered depressed scar and the slit is circular, i.e., neither horizontal nor vertical (the slit is transverse in ascites, and vertical in the presence of ovarian cyst).

Causes of *inverted and everted umbilicus* :

(A) **Everted**—Seen in any condition giving rise to increased intra-abdominal tension like ascites, ovarian cyst, pregnancy, umbilical hernia, hydramnios, severe gaseous distension etc.

(B) **Inverted**—Normally in health and in obesity (in obesity, umbilicus is buried in fat).

Observations on skin over abdomen (inspection) :

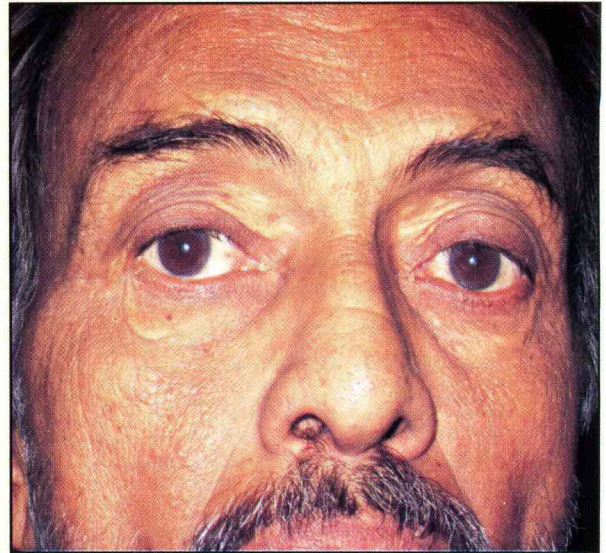
1. Moles, seborrhoeic warts, angiomas (considered normal changes).



Diagonal ear lobe crease – a strong morphological feature of coronary artery disease



A patient having **deep jaundice** with signs of senilities, eg, **arcus senilis**, cataract and 'bags under the eyes'; few seborrhoeic warts are seen around the eyes



Xanthelasma (soft, yellowish, plaque-like lesion) around the eyes and cataract are seen in a patient with ischaemic heart disease; xanthelasma is regarded as a physical sign of underlying hyperlipidaemia



Bilateral **sabre tibia** in osteopetrosis (marble bone disease) – 'sabre' literally means sword with its convex curved blade



Scaphoid abdomen (sunken or boat-shaped) in advanced starvation; wasting of intercostal muscles (due to loss of subcutaneous fat) is also noted



Flaccid bullae of **pemphigus**; rupture of blisters / bullae resulted in denuded areas in places



Erythematous, sharply-defined, plaque-like, coin-shaped, scaly (in places) lesions of **psoriasis** in anterior surface of legs in a young female



Bed sores (trophic changes) with exposed deeper tissues are seen over buttock and inferior angle of right scapula in a paraplegic patient

2. Scars, haemorrhagic spots, hypo- or hyperpigmentation (e.g., linea nigra—midline pigmentation below umbilicus seen in pregnancy), bruising periumbilically (Cullen's sign) and in flanks (Grey Turner's sign) in acute pancreatitis, vesicles of herpes zoster (may produce abdominal pain), marking for paracentesis abdominis (benzene stain or cotton seal), parietal oedema, shininess etc.
3. Striae : **White** striae or striae albicantes (obese persons who lost weight rapidly, following pregnancy or relieving ascites); **purple** striae usually represent the rupture of subepidermal connective tissue, Cushing's syndrome or prolonged steroid therapy); broad silvery lines (striae gravidarum) are stretch marks following repeated pregnancies.

Signs of 'peritonitis' :

The bedside findings are :

- a) Inspection : Distension, absence of abdominal movements, absent visible peristalsis.
- b) Palpation : Tenderness, rebound tenderness, 'board-like' rigidity.
- c) Percussion : Loss of liver dullness (e.g., in perforation of hollow viscus); very tender abdomen (avoid percussion).
- d) Auscultation : Absence of bowel sounds.

Table 21 : Differentiation between transudate and exudate

Features Transudate	Exudate
1. Appearance Clear, thin; does not clot on standing	Turbid, straw-coloured, haemorrhagic, chylous, mucinous or purulent. Often clots on standing
2. Specific gravity <1.016	> 1.016
3. Proteins <2.5 g/dl	> 2.5 g/dl
4. Cells < 250/mm ³ (i.e., low).	> 250/mm ³ (i.e., high). Endothelial cells or mesothelial Lymphocytes, neutrophils cells, and occasionally or malignant cells lymphocytes are seen

* In pancreatic ascites, amylase activity in the fluid is >1000 U/L; and low glucose concentration is found in tuberculous and malignant peritonitis.

Causes of transudative and exudative ascites :

(A) Transudate— (B)

1. Cirrhosis of liver.
2. CCF.
3. Nephrotic syndrome.
4. Hypoproteinaemia with severe anaemia.
5. Protein-losing enteropathy.
6. Constrictive pericarditis.
7. Pericardial effusion.
8. Meig's syndrome.
9. Myxoedema.
10. IVC obstruction.

Exudate—

1. Peritonitis—
 - (i) Pyogenic
 - (ii) Tuberculous
 - (iii) Malignant
 - (iv) Spontaneous bacterial
2. Pancreatic ascites.
3. Budd-Chiari syndrome.
4. Lymphoma or leukaemias.
5. Rupture of hollow viscus.
6. Injury or obstruction to the thoracic duct.

* Myxoedema, Meig's syndrome and chylous ascites may be exudative.

What is chylous ascites ?

1. Chylous—Turbid, creamy or milky peritoneal fluid due to presence of lymph. It shows Sudan III-staining fat globules and contains a large amount of triglycerides. Ether dissolves the turbidity of chylous ascites.
2. Chyliform ascites—Turbid fluid due to large number of leucocytes, degenerated cells or tumour cells. Addition of alkali clears the turbidity.
3. Pseudochylous ascites—Turbid fluid due to increased amount of lecithin, globulin and calcium phosphate in the ascitic fluid.

Causes of chylous ascites :

1. Tuberculosis
2. Filariasis
3. Trauma to the abdomen
4. Intra-abdominal malignancy
5. Nephrotic syndrome
6. Congenital lymphangiectasia or obstruction to the thoracic duct (e.g., lymphoma).

Ascites with lymphadenopathy :

1. Disseminated tuberculosis,
2. Disseminated malignancy.
3. Lymphoma, and
4. ALL and blast crisis of CML.

Causes of purulent and haemorrhagic ascites :**(A) Purulent ascites—**

1. Pyogenic peritonitis.
2. Pyaemia and septicaemia.
3. Ruptured amoebic liver abscess.
4. Penetrating injury to the abdomen.
5. Rarely, from pelvic inflammatory disease.

(B) Haemorrhagic ascites—

1. Abdominal trauma or traumatic tap.
2. Intra-abdominal malignancy.
3. Tuberculous peritonitis.
4. Bleeding diathesis.
5. Acute haemorrhagic pancreatitis,
6. Thrombosis of mesenteric artery.

Rapidly developing ascites :

1. Malignant peritonitis.
2. Tuberculous peritonitis.
3. Budd-Chiari syndrome.
4. Spontaneous bacterial peritonitis.
5. Lymphoma and leukaemia.
6. Chylous ascites.

Hepatomegaly with ascites :

1. Cirrhosis with portal hypertension.
2. Malignancy of liver.
3. Congestive cardiac failure (CCF).
4. Constrictive pericarditis.
5. Disseminated tuberculosis.
6. Budd-Chiari syndrome.
7. Lymphoma.

Splenomegaly with ascites :

1. Cirrhosis of liver with portal hypertension.
2. Lymphoma.
3. Systemic lupus erythematosus (SLE).
4. Disseminated tuberculosis.
5. Acute leukaemia.

Possible causes of refractory ascites :

Medical therapy does not give relief in 10-20% cases of ascites. The conditions contributing to refractory ascites are :

1. Non-compliance (not following the adequate restriction of sodium)
2. Functional renal failure in cirrhosis of liver
3. Infection
4. Spontaneous bacterial peritonitis
5. Conversion to hepatoma
6. G. I. bleeding
7. NSAID (depress diuretic effect of frusemide and spironolactone)
8. Portal or hepatic vein thrombosis.
9. Superimposed cardiac or renal disease.

Management : insufficient response to dietary and diuretic regime indicates development of refractory ascites, and is managed by salt-free albumin infusion (25 g in 3 hours), large volume paracentesis abdominis, Le Veen shunt or side to side portocaval shunt, and liver transplantation.

Ascites formation in cirrhosis :

1. Hypoalbuminaemia (principal cause)—Decreases the plasma oncotic pressure.
2. Portal hypertension (only localises the fluid in the peritoneal cavity).
3. Secondary hyperaldosteronism.
4. Increased level of ADH.
5. Increased oozing of 'hepatic lymph' from the surface of liver ('weeping liver').

Recent theory — Cirrhotic patients are always in a state of peripheral arterial vasodilatation -> diminished arterial blood volume -> rise in renin, aldosterone, vasopressin and noradrenaline -> leads to renal vasoconstriction, and sodium plus water retention. Moreover, the combination of splanchnic arterial vasodilatation and portal hypertension -> alters intestinal capillary permeability -> promoting formation of ascites. Vasodilators are of intestinal origin (NO, substance P) and are produced in response to endotoxin and cytokines.

* It is important to remember that in the absence of concomitant hypoalbuminaemia, portal hypertension alone does not lead to ascites in cirrhosis.

Important physical findings for aetiological diagnosis in ascites :

1. Virchow's gland—Gastrointestinal malignancy; gland in other areas in lymphoma.
2. Skin changes of hepato-cellular failure—Usually from cirrhosis of liver.
3. Engorged and pulsatile neck vein—CCF, pericardial effusion, constrictive pericarditis.
4. Prominent abdominal veins—Portal hypertension.
5. Sister Marie Joseph's nodule around umbilicus—Intra-abdominal malignancy.
6. Non-pitting oedema in feet—Myxoedema, filariasis.
7. Splenomegaly—Favours the diagnosis of portal hypertension (cirrhosis of liver) mainly; also found in lymphoma, leukaemias, Budd-Chiari syndrome.

8. Hepatomegaly—Firm liver indicates cirrhosis, and a hard, tender liver indicates malignancy.
9. Tenderness in ascites—Peritonitis (tuberculous, pyogenic, malignant ascites or SBP).
10. Rectal and vaginal examination—For any rectal or pelvic malignancy, haemorrhoids.

Ascitic fluid in cirrhosis and tuberculous peritonitis :

(A) Cirrhosis of liver—it is transudate in nature.

1. CLEAR, straw-coloured, light green or bile-stained fluid.
2. Specific gravity < 1.016.
3. Protein content < 2.5 g/dl.
4. Cells < 250 cells/mm³, mostly endothelial; Gram stain and culture—negative.
5. Serum-ascites albumin gradient (SAAG) > 1.1g/dl.

(B) Tuberculous peritonitis—it is exudate in nature.

1. Straw-coloured, haemorrhagic, mucinous or chylous fluid.
2. Specific gravity > 1.016.
3. Protein content > 2.5 g/dl.
4. RBC may be present.
5. Cells—1000/mm³, > 70% lymphocytes.
6. Serum-ascites albumin gradient (SAAG) < 1.1 g/dl.
7. Centrifused deposits stained by Ziehl-Neelsen's method may show tubercle bacilli. The tubercle bacilli may be cultured from ascitic fluid. The adenosine deaminase level in fluid is increased. The diagnosis is confirmed by peritoneal biopsy under laparoscopy.

What is 'serum-ascites albumin gradient' (SAAG) ?

- (A) It is the difference of albumin between serum and ascitic fluid. If the 'serum albumin' minus 'ascitic fluid albumin' (gradient) is equal to or greater than 1.1 g/dl, it suggests underlying portal hypertension. Other than cirrhosis of liver SAAG > 1.1 is a feature of CCF, Budd-Chiari syndrome, portal vein thrombosis, myxoedema and constrictive pericarditis.
- (B) If the gradient is less than 1.1 g/dl, it suggests malignant ascites, tuberculous ascites, pyogenic peritonitis, pancreatic ascites or nephrotic syndrome.

* SAAG is based on oncotic-hydrostatic balance and the gradient directly correlates with portal pressure. SAAG > 1.1 and ascitic protein < 2.5 g/dl is usually suggestive of portal hypertension.

Indications of abdominal paracentesis :

Paracentesis literally means removal of fluid. Actually this is divided into **diagnostic (No. 1 below) and therapeutic (No. 2, 3, 4, 5 below) indications**. As a whole, they are :

1. Diagnostic paracentesis (approximately 20-50 ml fluid is required).
2. Marked abdominal discomfort or cardio-respiratory embarrassment.
3. Refractory to medical therapy.
4. Danger of strangulation of umbilical hernia, if present.
5. Paracentesis may allow better abdominal examination, needle biopsy of liver, scanning or USG.

Dangers of abdominal paracentesis :

1. Sudden cardio-respiratory distress or shock in sudden and rapid withdrawal of large amount of fluid (if appears during the paracentesis, immediately stop tapping the fluid).
2. Introduction of infection (peritonitis).
3. Precipitation of hepatic coma (the compressed porto-caval shunts open up and nitrogenous materials reach the brain by-passing the liver).
4. Perforation of hollow viscus.
5. Protein depletion (5 litre of ascitic fluid may contain 50-100 g of protein).
6. Constant oozing of fluid (specially in tense ascites or malignant ascites).

Complications of ascites :

1. Collection of fluid in the pleural sac (commonly right-sided)
2. Spontaneous bacterial peritonitis.
3. Hernia (umbilical, inguinal, femoral); divarication of recti in tense and long-standing ascites.
4. Mesenteric venous thrombosis.
5. Functional renal failure.

How USG of abdomen helps in the diagnosis of aetiology of ascites ?

USG diagnoses the presence of free fluid in the abdomen and also detects the,

1. Presence of any mass, enlarged pre- or para-aortic lymph nodes.

2. Splenic enlargement.
3. Liver—Heterogenous echo-pattern (cirrhosis of liver) or diagnose malignancy of liver; oedema or inflammation of pancreas in acute pancreatitis.
4. Increased size of portal vein (>13 mm) in portal hypertension; presence of collaterals.
5. Any cyst—There may be malignant ascites with malignant ovarian cyst.
6. Enlargement of caudate lobe—Seen in Budd-Chiari syndrome.

Investigations you like to perform in a case of ascites :

1. Routine blood examinations—Hb, TC, DC, including ESR; neutrophilia indicates infection.
 2. Urine examination—High albumin in urine is found in nephrotic syndrome.
 3. Stool for occult blood—For abdominal malignancy and cirrhosis of liver.
 4. Serum cholesterol—It is increased in nephrotic syndrome and myxoedema; diminished in cirrhosis of liver.
 5. Plasma proteins—Low albumin level is seen in nephrotic syndrome, cirrhosis of liver, hypoproteinaemia with anaemia and protein-losing enteropathy.
 6. X-ray of the abdomen in erect posture—Ground-glass opacity i.e., diffuse abdominal haziness with loss of psoas margins in ascites (minimally requires 800 ml fluid); not much informative.
 7. **Ultrasonography of the abdomen is the best means to confirm ascites** / CT scan of the abdomen provides similar information. USG can detect as little as 30 ml of ascitic fluid.
 8. Examination of ascitic fluid (physical, biochemical, cytological and bacteriological study)—Gram's stain, acid-fast stains and culture should be performed. SAAG should be determined.
 9. Biopsy of Virchow's gland or any palpable lymph node — Informative in tuberculosis, lymphoma and malignancy.
 10. Tests of portal hypertension—See the section on 'Cirrhosis of liver.'
 11. Liver function tests (bilirubin, AST, ALT, alkaline phosphatase, albumin, globulin etc).
 12. Laparoscopy (peritoneoscopy)—May reveal peritoneal deposits of tuberculosis or malignancy.
 13. Liver biopsy or needle biopsy of peritoneum (in exudative ascites specially).
 14. Laparotomy.
- * Routine investigations like chest X-ray (pulmonary tuberculosis, cardiomegaly from CCF), ECG and echocardiography (CCF, pericardial effusion, constrictive pericarditis) should also be performed.

How to manage a case of ascites ?

Maximally, 500-700 ml fluid can be mobilised in 24 hours by the following :

1. Rest in bed mobilises ascitic fluid and helps in diuresis as renal perfusion increases in recumbency.
2. Diet—Salt restricted diet is given.
In severe cases, sodium in the diet should be less than 10 meq/day, i.e., 200 mg (in an average case 22 meq or 440 mg of sodium is allowed daily i.e., no extra salt, only minimum addition of salt during cooking is allowed). Total calories given daily are 2000-2200. One must be cautious while giving high protein diet if the patient shows features of neuro-psychiatric manifestations (from cirrhosis of liver). 1 g/kg of body weight of protein (only vegetable protein) is allowed unless there is any evidence of hepatic pre-coma.
3. Diuretics : they are introduced in a step-wise manner—
 - (i) Spironolactone (drug of choice)—100-400 mg/day in divided doses. Start with 25 mg thrice daily and gradually increase the dose (remember, it may produce hyperkalaemia).
 - (ii) Furosemide—40-80 mg/ day, if good response is not obtained by spironolactone alone.
 - (iii) Combination of i) and ii)
 - (iv) Other drugs like amiloride, bumetanide (1 mg/day), torasemide (5-10 mg/day) maybe tried.
4. Water intake—Restrict the intake to 1-1.5 litre daily.
5. Salt-free albumin infusion may be helpful.
6. Paracentesis abdominis—Where indicated (now-a-days, large volume paracentesis is done in cirrhosis of liver; 3-5 litre fluid is aspirated in one sitting with 6-8 g/L of salt-free albumin infusion). Dextran 8 g/L of ascitic fluid or haemaccel 125 ml/L of ascitic fluid may be infused instead of albumin to maintain the circulation.
7. Antituberculosis treatment is started in tuberculous peritonitis.
8. Treatment of other aetiologies—CCF, pericardial effusion, myxoedema, nephrotic syndrome, malignancy etc.
9. Peritoneo-venous shunt—Ascitic fluid is drained directly into Internal jugular vein and is known as Le Vein shunt.

10. Transjugular intrahepatic portosystemic stent shunting (TIPSS) may relieve resistant ascites in cirrhosis — a stent is placed between portal vein and hepatic vein in liver under radiological control.

11. Liver transplantation—In patients with cirrhosis of liver refractory to medical treatment.

* **Guide to progress**—By weighing the patient (should not fall >1 kg /day if ascites and oedema are present, and should not fall >0.5 kg/day in ascites alone) with measuring the girth of abdomen daily (often unreliable due to gaseous distention) and checking the serum electrolytes twice weekly.

Case 32

ABDOMINAL LUMP

How do we divide the abdomen for clinical examination ?

Abdomen is conveniently divided by two horizontal and two vertical lines into **nine distinct areas**.

The **vertical lines** pass from the femoral artery below to cross the lower costal margins close to the tip of the ninth costal cartilage (or midinguinal point below to midclavicular point above). The two **horizontal lines** are:

- (i) Subcostal or transpyloric (upper one), and
- (ii) Inter-tubercular (lower one).

The subcostal line is formed by joining the tips of the tenth costal cartilages on either side, the transpyloric plane lies midway between suprasternal notch and symphysis pubis, and the inter-tubercular line is formed by joining the tubercles on the iliac crests.

The different areas with contents are (clockwise)—

1. Right hypochondrium (right lobe of liver, gall bladder, hepatic flexure of colon).
2. Epigastrium (left lobe of liver, stomach, transverse colon, lower end of oesophagus).
3. Left hypochondrium (fundus of stomach, spleen, tail of pancreas, splenic flexure of colon).
4. Left lumbar (left kidney and adrenal gland, left ureter, descending colon).
5. Left iliac fossa (part of descending and sigmoid colon, left ovary and fallopian tube).
6. Hypogastrium or suprapubic (urinary bladder, uterus, sigmoid colon, rectum).
7. Right iliac fossa (caecum, appendix, part of ascending colon, right ovary and fallopian tube).
8. Right lumbar (right kidney and adrenal gland, right ureter, ascending colon).
9. Umbilical area in the middle (aorta, IVC, part of stomach, duodenal loop, small intestine, head and body of pancreas, and mesentery).

What is midclavicular line (MCL) ?

It is a vertical line drawn from the centre of the clavicle i.e., *above*, the line is passing midway between the middle of the suprasternal notch and the acromioclavicular joint; the *lower* point is the midinguinal point (midpoint between anterior superior iliac spine and symphysis pubis).

In males, the MCL is approximately in the line with the nipple.

Normal shape or contour of the abdomen :

It is neither scaphoid nor distended. Some clinicians describe the shape as globular.

Causes of scaphoid (sunken) abdomen :

- | | |
|----------------------------|-----------------------------|
| 1. Severe dehydration. | 4. Tuberculous peritonitis. |
| 2. Emaciation or cachexia. | 5. Malignant peritonitis. |
| 3. Advanced starvation. | 6. Meningitis. |

* For causes of 'distension' of abdomen, read the section on 'Ascites' (page 269).

Points noted during superficial palpation of abdomen :

Prerequisites :

1. Warm your hands by rubbing (specially in winter) and trim your nails for satisfactory abdominal palpation. Stand on the right side of the patient.
2. Use the flat of the right hand (don't poke with fingers —wrist and forearm should be in same horizontal plane).
3. Prepare the patient with thigh flexed, face turned to the opposite side. The patient should breathe quietly. Exposure of abdomen is the same as done during palpation of liver.

4. Movements of the right hand should be gentle and come from slight flexion of metacarpophalangeal joints. The hand is held flat and 'moulded' to the abdominal wall.
5. Start from left iliac fossa (or from an area which is far away from site of pain) and move anti-clockwise to end in suprapubic and umbilical region. Try to examine the tender area (according to the patient) at the last.

Points to note :

1. Surface temperature.
2. Tenderness.
3. Parietal oedema.
4. Hyperaesthesia.
5. Consistency (normal elastic feel, muscle guard or rigidity).
6. Any localised lump.
7. Divarication of recti (detected by rising test).
8. Pulsation (transmitted or expansile).
9. To determine the direction of blood flow in prominent abdominal veins, and
10. Fluid thrill with girth of the abdomen at the level of the umbilicus.

Normal consistency of the abdomen :

Normally the abdomen is elastic in feel (doughy in tuberculous peritonitis); 'board-like' rigidity in peptic perforation, tetanus and black widow spider bite.

- Local guarding—tender, inflamed viscus underneath (e.g., acute cholecystitis) or localised peritonitis.
- Generalised guarding—due to anxiety.
- Rigidity—indicative of peritonitis.

How to describe the lump or mass present in the abdomen ?

Preliminary preparations of the patient are the same as done during palpation of liver. Palpation of a lump falls within the purview of 'deep palpation' of the abdomen. Following points are to be described:

1. Site.
2. Size.
3. Shape.
4. Overlying skin (whether red, tense, shiny or pigmented).
5. Consistency (soft, firm, hard or cystic).
6. Margin (sharp or rounded).
7. Surface (smooth or nodular).
8. Mobility.
9. Tenderness.
10. Movement with respiration.
11. Parietal or intra-abdominal.
12. Pulsatile or not.
13. Bimanually palpable or not; ballotable or not.
- (14. Lastly palpate the liver, spleen and other viscera areawise, and examine the deep tender spots (McBurney's point, duodenal point, renal angle, gall bladder point). Also examine the hernial sites, detect the presence of free fluid in the abdomen and do the P/R, P/V examinations].

An abdominal mass is palpable : what to do ?

So far as lump abdomen is concerned, the abdomen is a temple of surprise. First **aim** is to decide the organ of origin and secondly, the pathological nature of the lump.

1. **Intra-abdominal or parietal ?**
 - (i) Ask the patient to raise the shoulders from the bed with arms over the chest, and then press firmly against the forehead (**rising test**), or
 - (ii) Ask the patient to raise both extended legs from the bed (leg lifting test).
Now one has to observe whether the swelling becomes less visible or disappears (intra-abdominal), or becomes more prominent or remains same (parietal). Parietal swelling like lipoma, fibroma, neurofibroma or large sebaceous cyst are of surgical interest.
Now place the patient on **knee-elbow position**. An intra-abdominal swelling becomes more prominent but not a retroperitoneal one.
2. Next, note the **region** where the swelling is present and think of the anatomical organs which are normally present there. For example, a mass palpable in the right hypochondrium may arise from liver, gall bladder, right kidney or hepatic flexure of colon.

3. Now try to '**get above the swelling**' if the mass is in the upper abdomen and try to 'get below the swelling' If it is present in the lower abdomen. For example, in case of a hepatic, splenic, renal or gastric mass, one cannot 'get above' the swelling. Similarly one cannot 'get below' a lower abdominal mass if it arises from uterus, urinary bladder, or upper rectum.
4. **Size and shape**—Swelling arising from liver, spleen, kidney, uterus or urinary bladder do not produce any problem for diagnosis in majority of cases. But if the swelling arises from stomach, small or large gut, pancreas (retroperitoneal structure) or peritoneum, it may be very difficult to diagnose; a large mass growing from the above mentioned organs usually distort their normal anatomical configuration. Note the site, shape, size by proper deep palpation.
5. **Margin, surface, tenderness, consistency and pulsation**—Smooth, round, non-tender, regular and tense swelling is usually cystic and benign whereas a hard, irregular, nodular and tender swelling is likely to be malignant in nature. A solid, ill-defined very tender mass suggests an inflammatory lesion. Note the overlying skin (any ulcer, sinus, peau d'orange appearance, redness, pigmentation) and examine for any pulsation (transmitted or expansile, judged by "Two fingers' test") present in the swelling.
6. **Movement with respiration and mobility**—Place the hand over the lower margin of the swelling and ask the patient to take deep breath. If the swelling moves up and down with respiration, it is obviously an intra-abdominal swelling (never parietal). Swelling arising from fixed organs like liver, spleen, kidneys, gall bladder and distal stomach show downward movement with respiration, however one cannot move the structures with the hand. But the swelling arising from structures having a broad base (mesentery or peritoneum) can be made to move freely by hands but they show no movement with respiration e.g., tumour of small and large gut, mesenteric cyst, secondary deposits in greater omentum.
A totally fixed mass is usually of retroperitoneal origin, like the pancreatic mass. A side to side mobility is present in fibroid uterus which is not a feature of full bladder or swelling arising from ovary. An ovarian tumour with long pedicle is mobile in all directions.
7. **Bimanually palpable or ballot table**—Renal mass is always bimanually palpable and ballotable. Even rarely a posteriorly situated gall bladder or a mass in the postero-inferior part of right lobe of liver may be bimanually palpable.

* For palpation of viscera of pelvic origin, empty the urinary bladder.

** For a pulsatile mass, perform "Two finger's test"—Place two index fingers on two sides of the mass as far apart as the mass allows and observe whether the fingers are raised and separated (expansile pulsation due to aortic aneurysm), or only raised (transmitted pulsation due to mass in the abdomen).

D/D of cystic swellings in the abdomen :

1. Ovarian cyst—See the section on 'Ascites'.
2. Encysted ascites (commonly seen in a patient with abdominal tuberculosis).
3. Mesenteric cyst—It is a well defined cystic swelling in the abdomen, usually lying in the umbilical region. It *moves freely at right angles to the line of attachment of the mesentery* than along the line. The line of attachment of the mesentery is an oblique line starting 1 inch left from the midline and 1/2 inch below the transpyloric plane (above), and extending upto the right sacro-iliac joint (below) for 6 inches.
4. Pseudopancreatic cyst—The smooth and round cyst has no mobility and often feels very firm rather than cystic one. Fluctuation may be absent. It is usually placed centrally above the umbilicus. Past history of trauma or acute pain abdomen (i.e., acute pancreatitis) is very important clue to diagnosis.
5. Hepatic (hydatid) cyst—The mass is present predominantly in the right hypochondrium. There may be presence of '**hydatid thrill**' (place three fingers firmly over the swelling and percuss the middle finger, when an 'after-thrill' produced by displacement of daughter cysts is felt by the index and ring fingers).
6. Renal cyst and hydronephrosis—The swelling is bimanually palpable and ballotable.
7. Splenic cyst—Should be differentiated from left-sided renal lump (read the section on 'Hepatosplenomegaly'); rarely bimanually palpable.
8. Choledochal cyst—Infant or an adult patient; commonly suffers from intermittent jaundice, fever and colicky pain in right upper abdomen. The mass feels cystic and is located in right hypochondrium.
9. Hydramnios—Associated with pregnancy.
10. FULL BLADDER—Last but not the least (read the section on 'Ascites' and see page 278).

Other important physical examination in 'lump abdomen' :

1. Anaemia.
2. Jaundice.

3. Assessment of nutrition.
4. Lymph nodes (specially Virchow's gland).
5. Pedal oedema. Any free fluid present in abdomen?
6. Temperature.
7. Examination of testes (one should not forget to palpate both testes).
8. Auscultation of the abdomen for signs of intestinal obstruction.
9. Palpation of other abdominal viscera.
10. Examination of CVS, respiratory, nervous and renal system.
11. P/RorP/V examinations.

Mass or lump in the right iliac fossa :

1. Ileocaecal tuberculosis.
2. Amoebic typhlitis (inflammation of caecum).
3. Appendicular lump.
4. Carcinoma of caecum or ascending colon.
5. Tubo-ovarian mass.
6. Crohn's disease.
7. Impaction of roundworms.
8. Intussusception.
9. Dropped or unascended right kidney.
10. Lymphoma.
11. Carcinoid syndrome.
12. iliac aneurysm.
13. Malignant undescended testicle.
14. Psoas abscess.

Mass or lump in the left iliac fossa :

1. Amoebic colitis.
2. Carcinoma of colon.
3. Diverticulitis.
4. Tubo-ovarian mass.
5. Colon loaded with stool (pits on pressure).
6. Dropped or unascended left kidney.
7. Lymphoma, psoas abscess.

Discharging 'sinuses' over abdomen :

2. Faecal fistula (post-operative).
3. Tuberculosis of the intestine.
4. Intra-abdominal malignancy.
6. Crohn's disease.
5. Scrofuloderma.
6. Actinomycosis.

* **Skin nodules** over abdomen : neurofibroma, lipoma, malignant deposits (e.g., sister Marie Joseph's nodule around umbilicus), calcinosis, sarcoidosis.

Classification of abdominal tuberculosis :

1. Tuberculosis of gastro-intestinal tract :
 - a) Ulcerative type (produces malabsorption).
 - b) Hyperplastic type (produces gut obstruction).
 - c) Ulcero-hyperplastic type.
2. Mesenteric lymph node tuberculosis (\pm tuberculosis of the mesentery).
3. Tuberculous peritonitis :
 - a) Plastic type.
 - b) Ascitic or exudative type; or encysted ascitic type (loculated).
 - c) Fibrous or adhesive type.
- [4. Tuberculosis of other abdominal solid organs like liver, pancreas, spleen etc.]

* When the exudate is small and more fibrinous, it binds the intestine, mesentery with or without involvement of lymph nodes, and is known as "tabes mesenteries".

'Lymph node lumps' in the abdomen :

These lumps feel variegated (firm to hard), surface may be nodular (due to individual large lymph nodes), occasionally tender (haemorrhage within) and may be present anywhere in the abdomen but in majority of cases they are present in epigastrium, umbilical area and right iliac fossa. They show restricted movement with respiration and there is absence of mobility. The common causes are,

1. Lymphoma (commonly pre- and para-aortic nodes).
2. Mesenteric lymph node tuberculosis.
3. Metastasis from carcinoma of neighbouring organs.
4. Metastasis from carcinoma of testes (always palpate the testes).
5. Filariasis (causes retroperitoneal lymphadenitis)—rare.

Cystic swelling in hypogastrium : is it urinary bladder ?

Following points should be noted carefully in a full bladder :

- (A) Midline oval swelling above symphysis pubis which may reach upto umbilicus.

SB) Palpation—Normally the urinary bladder is not palpable, it feels firm or elastic in retention of urine. Application of pressure induces pain and desire for micturition. Side to side movement of

the mass is not possible. In distension, upper and lateral borders are palpable but the lower border of the swelling can not be reached.

- (C) Percussion—It is done from above downwards and the swelling is dull on percussion.
- (D) Confirmation—When a simple rubber catheter is introduced through the urethra, urine comes out and the swelling disappears.

* D/D are gravid uterus (firm, side to side mobility, vaginal signs +), ovarian cyst (not truly midline, right- or left-sided), and fibroid uterus (firmer, often knobbly, vaginal signs +).

Common causes of palpable kidney :

(A) Unilateral:

1. Dropped kidney (can be pushed to its normal position).
2. Hydro- or pyonephrosis.
3. Wilms' tumour.
4. Hypernephroma.
5. Large cyst (solitary) in kidney.
6. Compensatory hypertrophy (other kidney damaged).

(B) Bilateral:

1. Polycystic kidney (irregular surface).
2. Bilateral hydronephrosis.
3. Bilateral dropped kidney.
4. Diabetic nephropathy in early stage.
5. Amyloidosis.
6. Scleroderma.
7. Acromegaly.

How to palpate the kidneys ?

The lower pole of right kidney is commonly palpable in thin patients for obvious reasons. The right kidney is palpated from right side though the left kidney may be palpated from either side. The method of palpation goes like this :

1. Preliminary preparations of the patient are the same as done during palpation of liver. Always sit on a stool for palpation of kidneys.
2. To palpate the right kidney, place the right hand horizontally in the right lumbar region anteriorly and the left hand is placed posteriorly in the right loin region to trap the kidney bimanually with inspiration (bimanual palpation).
3. Push the right hand in a backward, upward and inward direction, and ask the patient to take deep inspiration. A firm mass may be felt in between the two hands (if kidney is enlarged).
4. Next, a sharp tap is given by the left hand placed in the loin region. The anteriorly placed right hand now feels the kidney, and the kidney then falls back (by gravity) on the posterior abdominal wall which is felt again by the left hand. This is 'ballotement' (*confirmatory for kidney lump*). Ballotement demonstrates the mobility of the kidney.
5. The left kidney is then palpated by placing the right hand anteriorly in the left lumbar region and the left hand posteriorly in the left loin (from right side); place left hand anteriorly and right hand posteriorly in palpation of left kidney from left side.

Points to note in a renal lump :

1. Site
2. Size.
3. Shape (ovoid normally).
4. Consistency (resilient or firm in feel).
5. Margins (rounded).
6. Surface (normally smooth surface; irregular in polycystic kidney).
7. Tenderness.
8. Movement with respiration (normally kidney shows slight movement with respiration).
9. Whether bimanually palpable and ballotable.
10. Tenderness over the renal angle—Patient will be seated, and the angle formed by the 12th rib and lateral border of erector spinae muscle is pressed by the ball of the thumb or thrust given by ulnar aspect of closed fist—"Murphy's kidney punch". The test is done on both sides.
11. In left-sided renal lump—Examine for band of colonic resonance over the lump (by percussion).

* Renal angle is tender in acute pyelonephritis, perinephric abscess, nephrolithiasis, tuberculosis of the kidney. *Remember, a kidney lump is bimanually palpable and ballotable.* The kidney is ballotable because it is a posterior abdominal organ.

** A transplanted kidney is diagnosed by a) (transplanted) kidney in right or left iliac fossa, b) arterio-venous fistula in forearm (i.e., vascular access for haemodialysis), and c) laparotomy scar.

D/D of renal lump :

- | | |
|---------------------------------------|---|
| 1. Enlarged spleen. | 4 |
| 2. Carcinoma of bowel. | 5 |
| 3. Carcinoma or cyst of the pancreas. | 6 |

How to palpate the gall bladder ?

The gall bladder is palpated in the same way as the palpation of liver i.e., start palpation from right iliac fossa and move towards right hypochondrium. Normal gall bladder is not palpable. In pathological conditions, it is palpable just lateral to right rectus abdominis muscle near the tip of ninth costal cartilage. When palpable it feels firm, smooth and globular. When there is hepatomegaly, an enlarged gall bladder may be palpable in right lumbar or right iliac fossa. The comparative mobility on respiration and rounded borders differentiate it from right kidney lump.

Causes of palpable gall bladder :

1. Mucocoele/empyema of the gall bladder (firm and regular swelling).
2. Carcinoma of the head of pancreas (firm and globular swelling).
3. Carcinoma of the gall bladder (stony, hard, irregular swelling).
4. Carcinoma of the common bile duct (firm and globular swelling).

* With jaundice : 2, 3 and 4; without jaundice : 1.

D/D of gall bladder lump :

1. Hepatomegaly (specially, Riedel's lobe of liver).
2. Right-sided kidney lump or suprarenal tumour.
3. Carcinoma of the hepatic flexure of colon.
4. Carcinoma of the pyloric region of stomach.

Case 33**HEPATOSPLENOMEGALY****Common causes of hepatosplenomegaly :**

- | | |
|-------------------------------|--|
| 1. Chronic malaria. | 9. Polycythemia vera. |
| 2. Chronic kala-azar. | 10. Collagen vascular diseases like SLE. |
| 3. Haemolytic anaemia | 11. Tropical splenomegaly syndrome. |
| 4. Cirrhosis of liver. | 12. Myelofibrosis or myeloid metaplasia. |
| 5. Chronic myeloid leukaemia. | 13. Felty's syndrome. |
| 6. Lymphoma. | 14. Miliary tuberculosis or disseminated tuberculosis. |
| 7. Acute viral hepatitis. | |
| 8. Enteric fever. | 15. Amyloidosis. |

* ALL, AML, chronic lymphatic leukaemia, sarcoidosis, brucellosis, storage disorders (Gaucher's disease and glycogen storage disease) and polycystic disease are other important causes.

What is your diagnosis ?

1. Give the provisional diagnosis (aetiological e.g., it is a case of lymphoma), or say,
2. It is a case of hepatosplenomegaly probably due to chronic malaria, or say,
3. It is a case of hepatosplenomegaly for discussion (if aetiological diagnosis is not possible).

Different methods for palpation of liver :**During palpation of abdomen one must follow (mandatory) the undermentioned rules :**

1. Always stand on the right side of the patient (as mostly we are right-handed).
2. Flex the thighs (to relax the abdominal musculature).
3. Turn the patient's face to the left (so that patient can not breathe on your face).
4. Ask the patient to breathe deeply but regularly with open mouth (liver, spleen, gall bladder and kidneys move with respiration).
5. Hands of the patient will lie by the side of his/her trunk (helps to relax the abdomen).

Remember, the patient should lie flat comfortably on his back, on a firm mattress with the head supported by a pillow. There should be good light for the purpose of examination. In the winter season,

the hands of the examiner should be made warm by rubbing together vigorously. The patient should be relaxed and reassured. A gentle and unhurried palpation with care of the painful sites gain confidence of the patient and helps in extraction of maximal informations.

Exposure of abdomen from the xiphisternum to upper thigh (in males), or to just above the inguinal ligament (in females) is usually carried out in *inspection* of abdomen.

* Exposure of abdomen (for *palpation*) :

Above — The xiphisternum.

Below — Just above the inguinal ligament (do not embarrass the patient by exposing genitalia).

There are three techniques for palpation of liver. **Most of the clinicians in India are well conversant with the conventional method.**

I. Conventional method—

Place the flat of right palm firmly over the right iliac fossa and press the hand inwards and upwards. The HAND IS PLACED PARALLEL TO THE ARBITRARY LOWER BORDER OF LIVER (or the right subcostal margin) lateral to the right rectus abdominis muscle. At the height of inspiration press the fingers firmly inwards and upwards when the radial border of the right index finger will slip over the lower border of the liver, if it is palpable. At the phase of expiration, place the right palm on a higher level, by 1 cm at a time, over the anterior abdominal wall until you reach the right lower costal margin or detect the edge of liver. *Now palpate the epigastrium for the left lobe of liver (move the hand from right hypochondrium to midline).*

II. Preferred method (according to few clinicians) —

Place BOTH HANDS side by side flat on the anterior abdominal wall in the right hypochondriac region lateral to the right rectus abdominis muscle with the fingers pointing upwards. If any resistance is felt. MOVE THE HANDS FURTHER DOWNWARDS UNTIL THE RESISTANCE DISAPPEARS. The patient is then asked to inspire deeply, and at the height of inspiration press the fingers upwards and inwards to confirm the edge of liver. The manoeuvre is repeated from lateral to medial side in search of the lower border of liver. When the hand is moved downwards, the loss of resistance demarcates the lower border of liver.

III. Alternative method (according to few clinicians) —

The right hand is placed flat in the right iliac fossa WITH THE FINGERS DIRECTING UPWARDS, lateral to right rectus muscle (the finger tips lie parallel to rectus sheath). Now press the hand firmly upwards and inwards (index and middle one are the sensing fingers). At the height of inspiration, try to release the inward pressure while maintaining the upward pressure; with the inspiration, the tips of fingers will slip over the margin of the liver, if palpable. Now palpate the liver for surface, consistency etc as a routine. This manoeuvre is less sensitive than previous two methods.

* Dipping method is always applied in the presence of ascites.

** During palpation of liver, don't press patient's shoulder or bed by your left hand (a common mistake). Let it hang normally by the side of your body.

Points to note if liver is palpable :

1. Degree of enlargement—Expressed by centimetres / inches (preferable) or in number of examiner's fingers placed between the right costal margin and the lower border of the palpable liver at right MCL. Measurement is taken during natural expiration, preferably by a ruler. Some clinicians prefer to describe 'liver span' in this heading (see page 283).
2. Consistency—Soft, firm or hard.
3. Tenderness—Tender or non-tender.
4. Surface—Smooth or irregular (finely irregular or coarsely irregular).
5. Margin or border—Sharp or rounded. Usually a soft liver has rounded margin, and firm or hard liver has a sharp margin. The margin may be irregular in cirrhosis of liver.
6. Movement with respiration—Liver always moves 1 to 3 cm downwards with deep inspiration.
7. Left lobe enlarged or not (examine in the midline in the epigastrium between xiphoid process and umbilicus, and measure in mid-sternal line).
8. Any pulsation—Pulsatile or not (see below).
9. Upper border of liver dullness (normal or shifted)—Percuss the right side of chest from above downwards along the right MCL. Normally the upper border of liver dullness is found in right 5th ICS at right MCL. For further details read the section on Hydropneumothorax'.
10. Palpable hepatic rub.
11. Place the stethoscope over the liver and auscultate carefully for any hepatic bruit or hepatic rub.

* *Right lobe* of the liver is palpated by keeping the hand lateral to the right rectus abdominis muscle while the left lobe is examined in the midline. Caudate lobe enlargement (e.g., in Budd-Chiari syndrome) is also appreciated in epigastrium.

** During palpation, observe the facial grimace of the patient (for tenderness).

How to examine for pulsatile liver ?

1. Stand on the right side of the patient. While the patient is sitting in a chair (preferably), place your right palm over the liver (or right hypochondrium) and the left palm over the back, just opposite to the right palm (bimanual palpation).
2. Ask the patient to hold his breath after taking deep inspiration*.
3. Look from the side and observe the separation of the hands along with expansile pulsation of the liver, if there is any, and appreciate the pulsation.

Remember, the most common cause of pulsatile liver is CCF (produces functional tricuspid incompetence - TI). Thus, while examining the pulsatile liver, always look for engorged and pulsatile neck veins, and bipedal oedema (indirect evidence of CCF). Common causes of pulsatile liver are :

1. CCF (functional TI)
2. Organic TI (systolic pulsation).
3. Tricuspid stenosis (presystolic pulsation).
4. Haemangioma of liver or arteriovenous fistula in liver.
5. Transmitted epigastric pulsation from RVH.

* Inspiration helps in descent of liver and increases venous return to right heart.

Causes of tender liver :

- | | |
|---------------------------------------|------------------------------------|
| 1. Acute viral hepatitis. | 5. Drug-induced hepatitis. |
| 2. Congestive cardiac failure. | 6. Infected hydatid cyst of liver. |
| 3. Carcinoma of liver. | 7. Budd-Chiari syndrome (acute). |
| 4. Pyogenic or amoebic liver abscess. | 8. Cholangio-hepatitis. |

* Inflammation (perihepatitis) or stretching of the Glisson's capsule makes the liver painful and tender.

** Congestive hepatomegaly due to any cause (see below) makes the liver tender.

In an enlarged and tender liver, what else you will examine?

1. Jaundice (acute viral hepatitis).
2. Engorged and pulsatile neck veins, dependent bipedal oedema and CVS (for CCF).
3. Intercostal oedema as well as tenderness in right lower chest (liver abscess).

Causes of left lobe enlargement of liver :

1. Amoebic liver abscess.
2. Hepatoma.
3. Metastasis in liver.
4. Gumma of liver (not seen now-a-days)—hepar lobatum.

Causes of soft, firm and hard liver (alteration in consistency) :

(A) Soft liver [feels like lips]-

1. **Congestive cardiac failure.**
2. Acute viral hepatitis.
3. Fatty liver.
4. Acute malaria.
5. Viscerotropic normal liver.
6. Extrahepatic obstruction (EHO).

(B) Firm liver [feels like tip of the nose]~

1. **Cirrhosis of liver.**
2. Chronic malaria.
3. Chronic kala-azar.
4. Hepatic amoebiasis.
5. Lymphoma.
6. Chronic congestive cardiac failure (nutmeg liver).

(C) Hard liver feels like forehead-

1. **Hepatoma (hepato-cellular carcinoma) j Rock-hard liver**
2. **Metastasis in liver**
3. Chronic myeloid leukaemia.

* 'Congestive hepatomegaly' is commonly found in CCF, constrictive pericarditis, dilated (congestive) cardiomyopathy and Budd-Chiari syndrome.

Liver enlargement with irregular surface :

- (A) **Finely irregular** (granular surface)—Portal cirrhosis.
- (B) **Grossly or coarsely irregular** (nodular liver)—
 1. Postnecrotic cirrhosis (macronodular).
 2. Metastasis in liver (large nodules with umbilication due to central softening).
 3. Hepatoma.
 4. Hepatic cysts (congenital, hydatid).
 5. Multiple liver abscess.
 6. Gumma of liver.

* No. 1, 2, 4 and 5 produce 'knobbly liver'.

Causes of hepatic bruit :

1. Hepato-cellular carcinoma (hepatoma).
2. Acute alcoholic hepatitis.
3. Haemangioma of liver.
4. Arteriovenous fistula in liver.

Causes of hepatic rub :

1. After recent biopsy of liver (transient).
2. Perihepatitis (e.g., liver abscess).
3. Carcinoma of liver (secondary or primary).

Degree of enlargement of liver :

Enlargement is expressed as :

- Mild enlargement (1 -2 fingers)
- Moderate enlargement (2-5 fingers)
- Massive or huge enlargement (> 5 fingers)

Causes of huge hepatomegaly :

1. Metastasis in liver or hepatoma.
2. Polycystic disease of liver.
3. Gross fatty change.
4. Amoebic liver abscess.
5. Chronic malaria or chronic kala-azar.
6. Amyloidosis of liver.

* Mild to moderate hepatomegaly occurs in malaria, kala-azar, enteric fever, miliary tuberculosis, leukaemias, fatty liver, cirrhosis (postnecrotic), congestive hepatomegaly, haemolytic anaemia, carcinoma, haemangioma and storage disorders (Gaucher's disease).

Hepatomegaly with jaundice :

1. Acute viral hepatitis.
2. Haemolytic anaemia.
3. Carcinoma of liver (secondary).
4. Cirrhosis of liver.
5. Cholangio-hepatitis.
6. Weil's disease.
7. Lymphoma.

Hepatomegaly with heart failure :

1. Right ventricular failure.
2. Constrictive pericarditis.
3. Haemochromatosis.
4. Alcoholic liver disease with cardiomyopathy
5. Rarely, amyloidosis.

What is the 'liver span'?

It is the vertical distance between the uppermost and lowermost points of hepatic dullness. It is detected by percussing the upper and lower borders of liver at the right MCL. The anterior liver span is usually 10-14 cm (mean 12 cm) in an adult in the right MCL and is related to patient's age, sex, body mass, height etc (females having liver span less than males). Serial measurement is helpful to detect shrinkage or enlargement. Liver span in children is much smaller.

* Liver span is increased in true enlargement of liver and not when it is displaced (e.g., palpable liver in downward displacement in emphysema).

Causes of rapid change in size of liver :

(A) Rapid enlargement —

1. Carcinoma of liver.
2. Amoebic liver abscess.

(B) Rapid decrease —

1. Fulminant hepatic failure.
2. Correction of CCF.
3. Cirrhosis of liver.
4. Relief of cholestatic jaundice.

Conditions erroneously suggest upward hepatic enlargement :

1. Pleural effusion (right).
2. Basal pneumonia (right)
3. Subdiaphragmatic effusion or abscess (right).

D/D of hepatomegaly :

1. Carcinoma of the stomach.
2. Malignancy in right kidney.
3. Carcinoma of the transverse colon or hepatic flexure of colon.
4. Omental mass (generally from tuberculosis or malignancy).
5. Large gall bladder lump.

What is Riedel's lobe of liver ?

It is a downward tongue-like projection of the right lobe of liver and is commonly found in females. It moves with respiration. It is usually mistaken with gall bladder or right kidney. It lacks the spherical contour of the distended gall bladder and always remains asymptomatic. It is a congenital variation and not a true accessory lobe of liver. Often ultrasonography is done for differentiation.

N.B. : Palpable liver may be due to downward displacement of the liver without any enlargement and may be seen in conditions like right-sided pleural effusion, emphysema, in normal children, thin and lean persons, or in severe thoracic deformity. So, **a palpable liver may not be an enlarged liver**. Never forget to palpate the left lobe of liver, to percuss the upper border of liver dullness and to palpate bimanually for hepatic pulsation. Auscultation over the liver (for hepatic bruit and rub) is optional and should be done if you are asked to 'examine the liver'.

Different methods for palpation of spleen :

There are several methods for palpation of spleen. Preliminary preparations of the patient are the same as done during palpation of liver.

- I. Standing on the right side of the patient, **place the flat of the left palm firmly over the left costal margin** posterolaterally and press it forward and medially, so as to enable a slightly enlarged soft spleen closer to the palpating right hand. Ask the patient to breathe in deeply and start palpating the spleen with the right hand. **Starting from the right iliac fossa**, move upwards and laterally towards the left hypochondrium, 1 cm at a time between each breath. If the spleen is not palpable, move the right hand more upwards with each inspiration until the fingertips reach under the left costal margin. Start well out to the left of costal margin and gradually move more medially. It is better to palpate the spleen with the fingertips but few clinicians prefer to use the radial border of right index finger.
- II. If spleen is not palpable (or in a just palpable spleen) by the method mentioned above, turn the patient to **right lateral decubitus position** and palpate the spleen by hooked fingers of right hand placed under the left costal arch or by the same palpatory method mentioned above (palm lying flat) beginning close to the left costal margin while the patient is breathing in and out deeply. Patient will flex his left hip and knee while in right lateral decubitus position. The examiner's left hand should remain over the lowermost rib cage posterolaterally on the left side as before.
- III. In case of a just palpable spleen, finally stand on the left side of the supine patient, facing the foot end of the bed. Palpate the spleen by **hooked fingers of left hand** below the left costal margin while the patient breathes in and out deeply (classical hooking method). Hooking method may be done from the left side in sitting position of the patient.

* **If the spleen is not palpable by method I, go for method II and then, for method III. Phases of respiration are very important in palpation of spleen.** While palpating spleen, do not be hasty and rash, rather show endurance—a just palpable spleen will definitely touch your fingers at the height of inspiration. Method I and II may be called as bimanual palpation.

N.B. : Dipping method is always applied in the presence of ascites.

Different methods for palpation of abdominal viscera :

There are four methods :

1. Conventional, direct or classical method e.g., palpation of liver.
2. Hooking method — classically used in palpation of spleen.
3. Dipping method — palpation in the presence of ascites.
4. Bimanual palpation — classically for the palpation of kidneys.

What is dipping method ?

It is difficult to palpate the abdominal viscera by direct method in the presence of significant ascites. While performing dipping method, mould the palpating right hand to the shape of the abdomen and then tap the abdomen sharply and quickly. The sudden and rapid displacement of fluid gives a tapping sensation over the surface of the enlarged liver or spleen which is comparable to patellar tap. The right hand is placed flat over the abdomen and **sharp tap is given by flexing the metacarpophalangeal joints suddenly**. One or two sudden thrust displaces the fluid and the displaced fluid pushes the organ forward towards the palpating fingers. Many a time full description of an organ is not possible by dipping method (one can go upto palpable or not, tender or non-tender, small or big lump). Preliminary preparations of the patient are the same as done during palpation of liver. It is better to start palpation from right iliac fossa by dipping method and proceeding anticlockwise.

While palpating the spleen by dipping method, it is better to place the left palm over the left costal margin as done in the conventional method.

How to percuss the upper border of splenic dullness ?

The anatomical position of the spleen is behind and below the left 9th, 10th and 11th rib, and its **long axis is lying along the direction of the left 10th rib**. Its antero-inferior end extends maximally upto the midaxillary line and the postero-superior end lies 4 cm lateral to the T₁₀ spine. The upper border of normal splenic dullness is present in left 9th rib and for this reason the 8th ICS (space above the 9th rib) in left midaxillary line is always resonant on percussion.

Percuss the left axilla from above downwards along the midaxillary line (either the patient lies supine or being seated while keeping the left hand over the head to expose the axilla) in the search of upper border of spleen.

If the same percussion is done in lowest intercostal spaces (like 8th or 9th ICS) along the anterior axillary line and the spaces reveal dull percussion note, even on full inspiration, it is said that splenomegaly is present (**Castell's method**).

* Remember, splenic enlargement first takes place in superior and posterior direction before it becomes palpable per abdomen (i.e., subcostally).

Percussion of the lower border of liver :

1. Start percussion from below upwards i.e., from right iliac fossa to right hypochondrium along the right MCL. &
2. It is a light percussion (as lower border of liver is not covered by any organ).
3. Place the pleximeter finger parallel to right subcostal margin and the line of percussion will be perpendicular to that margin.

Percussion of the upper border of liver :

1. Start percussion from above downwards in the right chest along the right MCL.
2. It is a heavy percussion (as upper border of liver lies under cover of the right lung).
3. Place the pleximeter finger in the right 2nd ICS parallel to arbitrary upper border of liver and the line of percussion will be perpendicular to that border.

* For shifting of upper border of liver dullness, read the section on 'Hydropneumothorax'.

Percussion of the lower border of right lung :

1. Start percussion from above downwards in the right chest along the right MCL.
2. It is a light percussion (as it is a superficial organ).
3. Place the pleximeter finger in the right 2nd ICS (parallel to arbitrary lower border of lung) and the line of percussion will be perpendicular to that border.

Points to note if spleen is palpable :

1. Degree of enlargement—Expressed by centimetres / inches (preferable) or in number of examiner's fingers placed below the left costal margin upto the most distal point of spleen, along the splenic axis (spleen becomes just palpable when it is enlarged to 2-3 times more than its normal size. **Spleen usually enlarges towards the right iliac fossa**. In children, spleen may enlarge vertically towards the left iliac fossa). The measurement is taken along the splenic axis from a point on left costal margin (where crossed by left MCL) to the furthest extent of the spleen. Measurement is taken during natural expiration. A spleen, which is palpable, is always pathological.
2. Splenic notch—Very characteristic and felt on its anterior or medial border.
3. Margin—Usually the margin is sharp.
4. Consistency—Soft, firm or hard.
5. Tenderness—Tender or non-tender.

6. Surface—Smooth or irregular.
7. Movement with respiration—Spleen always moves downwards and medially with respiration.
8. Fingers can not be insinuated between the left costal arch and the enlarged spleen. Spleen has a tendency to bulge orward.
9. Palpable splenic rub—Present or not (for its detection, the patient must breathe in and out deeply).
10. Auscultation of splenic rub.
11. Upper border of splenic dullness.

N.B. : No. 10 & 11 are optional and should be done if you are asked to 'examine the spleen'. Some clinicians prefer that a student should always percuss the spleen (D/D with renal swelling) for any band of colonic resonance, at the time of palpation of spleen.

* It is not clearly known, why the spleen in children enlarges vertically towards the left iliac fossa. Probably, it may be one of the reason that in a child other organs are not much enlarged (in comparison to adults) and thus the spleen makes its room vertically downwards towards the left iliac fossa.

** In the examination, do not delineate borders of spleen/liver by pen or pencil over patient's body.

Table 22 : Differentiation between enlarged spleen and renal lump

Spleen	Left kidney
1. Spleen enlarges downwards, forwards and towards the right iliac fossa, and moves well with respiration	1. Kidney enlarges towards the left iliac fossa and having restricted movement with respiration
2. <i>Notchfelt</i>	2. No notch felt
3. Sharp margin	3. Rounded margin
4. It is neither bimanually palpable nor ballottable	4. <i>Bimanually palpable and ballottable</i>
5. <i>Fingers cannot be insinuated between left costal margin and the enlarged spleen</i>	5. Fingers can be insinuated
6. Percussion note is always dull over spleen	6. <i>Band of colonic resonance anterior to enlarged kidney may be elicited</i>
7. Tendency to bulge forward	7. Tendency to bulge into the loin
8. Renal angle is non-tender	8. Renal angle may be tender
9. Cannot 'get above the swelling'	9. May 'get above the swelling' (e.g., in dropped kidney)
10. Can cross midline	10. Does not cross midline

* A massive splenomegaly may be bimanually palpable but never ballottable.

D/D of splenomegaly :

1. Enlarged left kidney.
2. Enlarged left lobe of liver.
3. Carcinoma of the stomach.
4. Carcinoma of the splenic flexure of colon.
5. Omental mass (generally from tuberculosis or malignancy).
6. Malignancy of the tail of pancreas.

Different degrees of enlargement of spleen :

(A) Mild (upto 2 fingers) or just palpable spleen ('spleen tip') —

- | | |
|---|--|
| 1. Acute malaria. | 7. Acute leukaemias. |
| 2. Acute kala-azar. | 8. Haemolytic anaemia. |
| 3. Enteric fever. | 9. Immune thrombocytopenic purpura (ITP) |
| 4. Acute viral hepatitis. | 10. Felty's syndrome. |
| 5. Subacute bacterial endocarditis (SBE). | 11. Infectious mononucleosis. |
| 6. Miliary tuberculosis. | 12. Weil's disease. |

(B) Moderate (2-5 fingers or upto the umbilicus)—

- | | |
|-------------------------|---|
| 1. Chronic malaria. | 5. Chronic lymphocytic leukaemia. |
| 2. Chronic kala-azar. | 6. Collagen vascular diseases like SLE. |
| 3. Portal hypertension. | 7. Congenital haemolytic anaemia, |
| 4. Lymphoma. | e.g., thalassaemia. |

(C) Huge or massive (> 5 fingers, or > 7 cm, or when the spleen crosses the umbilicus)—

1. Chronic kala-azar or chronic malaria.
2. Thalassaemia major.
3. Chronic myeloid leukaemia.
4. Tropical splenomegaly syndrome.
5. Polycythemia vera.
6. Portal hypertension with hypersplenism (in cirrhosis).
7. Extrahepatic portal vein obstruction (EHO).
8. Myelofibrosis or myelosclerosis.
9. Storage disorders (Gaucher's disease), amyloidosis.
10. Splenic cyst, tumour or abscess.

* Normal spleen weighs < 250 g; in massive splenomegaly, it weighs > 1000 g in adults.

** Remember, all the causes of massive splenomegaly were mild to moderate splenomegaly one day, and all moderate splenomegaly were mild in nature to start with.

*** Spleen may be palpable without having actual enlargement in tall and lean persons, emphysema (hyperinflated lung pushes spleen), visceroptosis and sometimes in children.

When do you search for splenic rub ?

Splenic rub is heard in the situations where the patient complains of acute pain in left upper quadrant of abdomen (often with radiation of pain to the tip of left shoulder) due to splenic infarction resulting in perisplenitis (with acute splenic tenderness). It is produced in conditions like,

1. Subacute bacterial endocarditis (SBE).
2. Chronic myeloid leukaemia.
3. Sick cell anaemia.
4. After splenic puncture (e.g., in diagnosis of chronic kala-azar).
5. Vasculitis.
6. Splenic torsion.

* Acute splenic tenderness indicates splenic infarction or abscess formation.

The characteristics of splenic rub are :

1. It is a scratchy to-and-fro sound heard with respiration i.e., during movement of the spleen.
2. If the patient holds his breath, the rub stops.
3. The diaphragm of stethoscope should be placed over the spleen or left lower chest.

It should be differentiated from left-sided pleural rub (spleen is always tender in splenic rub) which is often very difficult. Treatment is by rest and analgesia. Repeated splenic infarction may be an indication for splenectomy.

** Always auscultate for splenic rub over a hugely enlarged spleen. Splenic rub may be palpable.

*** Spleen may be mildly tender in acute malaria, infectious mononucleosis, SBE and enteric fever.

**** **Splenosis**—peritoneal seeding of splenic rupture (traumatic) fragments not connected to portal circulation, and giving rise to abdominal pain later (like endometriosis).

What do you mean by hypersplenism ?

It is the splenic hyperactivity with increased blood cell destruction. Diagnostic criteria are :

1. Splenomegaly (of any size).
2. Pancytopenia (anaemia, leucopenia and/or thrombocytopenia).
3. Normal or hypercellular bone marrow.
4. Reversibility by splenectomy.

Causes — Either primary (no underlying disease) or secondary hypersplenism (connective tissue disorders, lymphoma, myeloproliferative disorders, cirrhosis of liver or congestive splenomegaly).

Causes of 'hyposplenism' :

It means absence of spleen or nonfunctioning spleen. It is usually associated with :

1. Dextrocardia (spleen is absent congenitally or asplenia).

2. Sickle cell disease (due to repeated splenic infarcts leading to autosplenectomy).
3. Coeliac disease.
4. Dermatitis herpetiformis with enteropathy.
 5. Fanconi's anaemia (aplastic anaemia with hypoplasia of spleen, kidney, thumb, or radii).
6. Surgical removal.

* Splenectomised or hyposplenic patients are prone to be infected with capsulated organisms like pneumococcus, meningococcus, H. influenzae or E. coli, and thus vaccination against these organisms is recommended to them.

Myeloproliferative disorders :

Here, there is uncontrolled clonal proliferation of one or more of the cell lines in the bone marrow, and are classified as :

1. Chronic myeloid leukaemia.
2. Polycythemia vera.
3. Essential thrombocythemia.
4. Myelofibrosis (myeloid metaplasia).

Splenomegaly with jaundice :

1. Cirrhosis of liver.
2. Acute malaria (P. falciparum commonly).
3. Haemolytic anaemia (e.g., thalassaemia).
4. Lymphoma.
5. Acute viral hepatitis (uncommon).
6. Miliary tuberculosis.

Hepatosplenomegaly with lymphadenopathy:

1. Lymphoma.
2. Leukaemia (ALL and CLL).
3. Disseminated tuberculosis.
4. SLE.
5. Sarcoidosis.
6. Felty's syndrome.

Hepatosplenomegaly with pyrexia :

1. Malaria (acute).
2. Kala-azar (acute or chronic).
3. Enteric fever.
4. SBE.
5. Miliary tuberculosis
6. Acute viral hepatitis.
7. Acute leukaemias, CML.
8. Lymphoma.
9. Collagen vascular diseases.
10. Brucellosis.
11. Still's disease.

Reliable methods for measuring actual splenic size :

These are imaging procedures like,

1. Radionuclide scan,
2. Ultrasonography (USG),

If a maximal cephalocaudal diameter of spleen by these techniques (especially USG) crosses 13 cm, spleen is said to be enlarged.

Causes of rupture of spleen :

1. Trauma in normal spleen.
2. By trivial trauma in a diseased spleen. The pathological conditions responsible are infectious mononucleosis, acute and chronic leukaemias, myelofibrosis, congestive splenomegaly (portal hypertension due to cirrhosis of liver or Budd-Chiari syndrome).
3. Computerised tomography (CT), and
4. Nuclear magnetic resonance imaging (MRI).

Hepatosplenomegaly with ascites :

1. Cirrhosis with portal hypertension.
2. Lymphoma.
3. Disseminated tuberculosis.
4. Acute leukaemia.
5. Acute viral hepatitis with hepato-cellular failure.
6. Systemic lupus erythematosus (SLE).

Splenomegaly with purpura or petechiae :

1. Acute leukaemia.
2. SBE.
3. Blast crisis of CML and CLL.
4. SLE.
5. ITP.
6. Felty's syndrome.
7. Infectious mononucleosis.
8. Amyloidosis.
9. AIDS.
10. Toxoplasmosis, CMV infection.
11. CML with blast crisis.
12. Kala-azar (African variety).

Hepatosplenomegaly with anaemia :

1. Chronic malaria.
2. Chronic kala-azar.
3. Haematological malignancies e.g., lymphomas and leukaemias.
4. Cirrhosis of liver.
5. Haemolytic anaemia (e.g., thalassaemia).
6. Severe iron deficiency anaemia.
7. Hypersplenism.
8. SBE.
9. SLE.
10. Still's disease.

Important physical signs in hepatosplenomegaly :

1. Anaemia (cirrhosis, thalassaemia).
2. Jaundice (cirrhosis of liver, thalassaemia).
3. Fades — Mongoloid or hepatic fades; butterfly rash in SLE.
4. Polycythemia (i.e, plethoric appearance).
5. Lymphadenopathy (lymphoma, lymphocytic leukaemia, infectious mononucleosis).
6. Spider naevi and palmar erythema (hepato-cellular failure from cirrhosis of liver).
7. Kayser-Fleischer ring in cornea (Wilson's disease).
8. Tremor (Wilson's disease).
9. Pigmentation (haemochromatosis).
10. Leg ulcers (congenital haemolytic anaemia).
11. Sternal tenderness (acute leukaemias and CML).
12. Haemorrhagic spots (acute leukaemias, SBE).
13. Fundoscopy—Roth spots (SBE), choroidal tubercle (miliary tuberculosis).

Investigations you like to perform in a case of hepatosplenomegaly :**(A) Blood — Hb, TC, DC, ESR and some special tests :**

- a) Leucocytosis—Pyogenic infections, polycythemia, leukaemia.
- b) Leucopenia—Malaria, kala-azar, enteric fever, Felty's syndrome.
- c) Pancytosis—Polycythemia.
- d) Pancytopenia—Hypersplenism.
- e) Reticulocytosis—Haemolytic anaemias.
- f) Thrombocytopenia—Leukaemia, hypersplenism.
- g) ESR—Increased in infective aetiologies, SLE, lymphoma, severe anaemia due to any cause; diminished in polycythemia and congestive cardiac failure.
- h) Blood culture—Often informative in SBE, enteric fever.
- i) Blood smear—For the diagnosis of malaria, kala-azar (buffy coat preparation), leukaemia (acute and chronic), hereditary spherocytosis, thalassaemia.
- j) Special tests—(i) Paul-Bunnell test - for infectious mononucleosis, (ii) Serum iron - increased in thalassaemia, haemochromatosis. (iii) Serum copper - low in Wilson's disease, (iv) Serum bilirubin - for cirrhosis with hepato-cellular failure, haemolytic anaemias, malaria, viral hepatitis. (v) Serum electrophoresis - increased HbF in thalassaemia. (vi) Congo red test for amyloidosis. (vii) Aldehyde test - for chronic kala-azar. (viii) LE cell and phenomenon - collagen vascular disease (not done routinely), (ix) Red cell survival time - decreased in hereditary spherocytosis, (x) Antinuclear factor - positive in SLE. (xi) Rose-Waaler test - positive in Felty's syndrome, (xii) Serum proteins - high globulin with low albumin is observed in cirrhosis of liver, chronic kala-azar, disseminated lupus, (xiii) Blood for HB_sAg. (xiv) Serology—for CMV, EBV, HIV, and VDRL test for syphilis.

(B) X-ray or radiology —

- a) Chest (PA view)—For miliary tuberculosis (miliary mottling), lymphoma or sarcoidosis (mediastinal widening), extramedullary haematopoiesis in thalassaemia.
- b) Bones —
 - (i) Mosaic-pattern seen in small bones of hand in patients with thalassaemia.
 - (ii) Increased bone density in myelofibrosis or myelosclerosis.
 - (iii) Expansion of lower end of long bones (Erlenmeyer flask)—Seen in Gaucher's disease.
- c) Skull— 'Hair on end' appearance in thalassaemia.
- d) Straight X-ray of abdomen—Not informative .

(C) Lymph node biopsy (excision biopsy is always preferred to FNAC because it delivers more tissue for diagnosis)—May be of value in tuberculosis, sarcoidosis and lymphoma.**(D) USG of liver, spleen, free fluid in abdomen and for any mass (pre- and para aortic lymph nodes)—**

- a) Hepatosplenomegaly is confirmed.
- b) Ascites may be present in portal hypertension, tuberculosis or lymphoma.
- c) Heterogenous echopattern of liver with splenomegaly and ascites—Portal hypertension due to cirrhosis of liver.

- d) Enlarged abdominal lymph nodes —Lymphoma or tuberculosis.
- (E) Liver biopsy—May be of value in cirrhosis of liver, lymphoma, haemochromatosis or amyloidosis.
- (F) Bone marrow examination—For leukaemia, lymphoma, hypersplenism, Gaucher's disease, polycythemia etc.
- (G) Skin test—
 - (1) Mantoux test—For tuberculosis (may be negative in lymphoma).
 - (li) Kveim test—For sarcoidosis (not done now-a-days).
- (H) Investigations for portal hypertension—See the section on 'Cirrhosis of liver'.
- (I) Splenic puncture—For the diagnosis of chronic kala-azar.
- (J) CT scan of whole abdomen may be done.
- (K) Lymphangiography—For lymphoma (for demonstration of involvement of retroperitoneal nodes).

D/D of hepatosplenomegaly :

I. Chronic malaria :

- a) Patient comes from an endemic zone with high rise of temperature associated with chill and rigor. Fever may come on alternate days periodically.
- b) Severe anaemia, mild jaundice may be present.
- c) Spleen—Moderate to massive enlargement, firm to hard and non-tender.
- d) Diagnosis is confirmed by demonstration of malaria parasites in the peripheral blood. There may be leucopenia.

II. Chronic kala-azar :

- a) Patient comes from an kala-azar endemic zone.
- b) Double rise of temperature (double quotidian) in 24 hours may be present (20% cases).
- c) Appetite is preserved inspite of fever and loss of body weight.
- d) Spleen—Massively enlarged, soft to firm (often a doughy feel) and non-tender (spleen of chronic kala-azar is softer than chronic malarial spleen).
- e) Liver—Moderate enlargement, firm and non-tender. Ratio of liver and spleen is 2 : 5.
- f) Alopecia, pigmentation.
- g) Diagnosis is made by neutropenia, positive aldehyde test, direct agglutination test (DAT), or by demonstration of L.D. bodies in buffy coat preparation of blood or bone marrow smear or splenic puncture.

m. Thalassaemia (Cooley's anaemia) :

- a) The patient is usually a child or young adult with positive family history.
- b) Stunted growth, typical mongoloid facies, massive splenomegaly with moderate hepatomegaly, severe anaemia, mild jaundice, leg ulcers over malleoli of foot.
- c) Diagnosis is done by,
 - (i) Microcytic hypochromic anaemia, reticulocytosis, anisocytosis, polikilocytosis, target cells,
 - (il) Haemoglobin electrophoresis (HbF >2%).
 - (iii) Radiological study of skull and small bones of hand.

* Acquired haemolytic anaemia (e.g., autoimmune) may produce hepatosplenomegaly.

IV. Cirrhosis of liver (postnecrotic) :

- a) Features of chronic liver disease e.g., dyspepsia, malaise, wasting, severe weakness. There may be past H/O jaundice.
- b) Symptoms and signs of hepato-cellular failure.
- c) Features of portal hypertension—Haematemesis, melaena, splenomegaly, venous prominences over abdomen, ascites.
- d) Mild to moderate hepatomegaly with moderate splenomegaly. Both are firm and non-tender.
- e) Presence of other stigmata of cirrhosis of liver.
- f) Diagnosis is done by demonstration of oesophageal varices, splenoportalvenography, liver biopsy.

V. Chronic myeloid leukaemia :

- a) The patient is around 30 to 80 years (peak - 55 years) with fever, dragging pain in the left hypochondrium and profound weakness. Weight loss and sweating are due to increased metabolic rate.
- b) Anaemia —Mild.

- c) Sternal tenderness —Present.
- d) Spleen —Hugely enlarged, hard with occasional splenic rub (may be tender).
- e) Liver —Mild to moderate enlargement.
- f) Diagnosis is confirmed by 1.5-2.5 lacs WBC/mm³ with myelocytes, metamyelocytes, promyelocytes and few myeloblasts.
- * (AML - mild splenomegaly; ALL - moderate splenomegaly
CML - huge splenomegaly; CLL - moderate splenomegaly]

VI. Lymphoma :

- a) Progressive painless enlargement of cervical, axillary or inguinal lymph nodes, fever, loss of weight, pruritus, weakness, drenching night sweats.
- b) Superior mediastinal syndrome may be present — Dyspnoea, engorged non-pulsatile neck vein, swelling of face, tortuous veins over chest wall.
- c) Spleen — Moderately enlarged, non-tender.
- d) Liver — Moderate enlargement.
- e) Anaemia, eosinophilia in blood may be present. The lymph node biopsy is confirmatory.

VII. Acute viral hepatitis :

- a) H/O jaundice preceded by anorexia, profound nausea, fever and occasional vomiting. Urine is yellow and stool may be of white coloured.
- b) There may be urticarial rash, arthralgia.
- c) Hepatosplenomegaly—Mild to moderate, tender hepatomegaly with mild splenomegaly (soft).
- d) Diagnosis is done by—
 - (i) High serum bilirubin, high SGPT (ALT), alkaline phosphatase (slight rise), prothrombin time estimation (may rise).
 - (ii) HB Ag, LM anti-HAV or anti-HEV, anti-HCV in blood.

VIII. Enteric fever :

- a) H/O rise of temperature in step-ladder pattern, frontal headache, constipation, epistaxis, rose spots in skin, relative bradycardia.
- b) Tongue—Red margins with central coating (angry-looking tongue).
- c) Spleen—Soft and tender just palpable spleen at the end of first week.
- d) Liver—Soft, non-tender, mild enlargement.
- e) Diagnosis is done by neutropenia, blood culture and Widal test.

IX. Polycythemia vera :

- a) Symptoms like headache, fullness in head, pruritus, peripheral vessel thrombosis (due to increased viscosity of blood), visual disturbances commonly in a middle aged male patient.
- b) Epistaxis, haematemesis or melaena, pruritus, peptic ulcer, peripheral vascular bleeding.
- c) Face is plethoric, conjunctivas are congested and palms are red.
- d) Spleen is enlarged in 75% cases and may be massive; hepatomegaly in 30% cases.
- e) Diagnosis is made by raised haematocrit, low ESR, hyperplasia of all marrow elements in bone marrow examination. Erythropoietin level is decreased.

X. Collagen vascular disease (SLE) :

- a) The patient is mostly a woman with fever, arthralgia, alopecia, oral ulcer, skin rash or lymphadenopathy.
- b) Renal, lung or cardiac involvement—Oliguria, pleurisy, pericarditis, dyspnoea (i.e., multisystem disease).
- c) Spleen—Usually there is moderately enlarged, firm, non-tender spleen,
- d) Diagnosed by high ESR, positive antinuclear factor, antibody to double-stranded DNA and anti-Sm antibody (SLE), renal biopsy etc.

XI. Tropical splenomegaly syndrome (previously known as Banti's syndrome) :

- a) The patient comes from a hyperendemic area with an exaggerated immune response to malaria. The disease is common in females, and has familial and racial predisposition. Adults have high malarial antibody titres with low parasitaemia.
- b) Spleen—Massive splenomegaly.
- c) Liver—Mild to moderate enlargement.

- d) Signs of portal hypertension may be present without any evidence of cirrhosis of liver (due to increased portal blood flow).
- e) Chronic low grade haemolytic anaemia may be seen; low grade pyrexia.
- f) Diagnosed by increased level of I_gM, 'sinusoidal lymphocytosis' in liver biopsy (diagnostic).

* The modern nomenclature of the disease is '**hyperreactive malarial splenomegaly**'.

XII. Myelofibrosis or myelosclerosis :

- a) This may be primary, or develops from toxins, malignancy, lymphoma, irradiation (secondary).
- b) Spleen—Massive, tender spleen; splenic rub may be present.
- c) Hepatomegaly—Mild to moderate enlargement.
- d) Leucoerythroblastic blood picture with high platelet count.
- e) Bone marrow examination may not yield any result (dry tap) and often trephine biopsy is needed to demonstrate the fibrotic changes. X-ray shows ground-glass appearance (osteosclerosis) of all bones of axial skeleton.
- f) There may be signs of haemorrhage into the skin.

XIII. Felty's syndrome :

- a) It is a subgroup of rheumatoid arthritis with splenomegaly and neutropenia.
- b) Age of onset is around 50-70 years and more common in females.
- c) Features like weight loss, joint pain with deformity, lymphadenopathy, pigmentation, leg ulcers, vasculitis, chest infection, keratoconjunctivitis sicca, rheumatoid nodules may be present.
- d) Spleen — Mild splenomegaly.
- e) Diagnosis is done by anaemia, neutropenia, thrombocytopenia, and strongly positive rheumatoid factor.

XIV. Miliary tuberculosis :

- a) Gradual onset with vague ill-health, high fever with drenching sweats, pallor, loss of weight.
- b) Cough, breathlessness, anorexia, headache may be present.
- c) Tachycardia, paucity of signs in the chest (few crepitations in late stages).
- d) Mild, non-tender hepatomegaly with small palpable spleen.
- e) Signs of meningeal irritation may be found.
- f) Choroidal tubercle may be visible (25%) on ophthalmoscopy (diagnostic). Chest X-ray shows miliary mottling bilaterally. Leucocytosis is usually absent. Mantoux test may be negative. Sputum examination may or may not demonstrate AFB.

XV. Amyloidosis :

- a) Presence of long standing suppurative focus in the form of lung abscess, bronchiectasis, chronic osteomyelitis or may develop from tuberculosis, Crohn's disease, multiple myeloma, lymphoma, leprosy, rheumatoid arthritis.
- b) Visceromegaly—Macroglossia (tongue becomes stiff and firm to palpation), hepatosplenomegaly.
- c) Evidence of nephrotic syndrome—Albuminuria, hypoproteinaemia, anasarca and hypercholesterolaemia.
- d) Malabsorption, lymphadenopathy, peripheral neuropathy, cardiomyopathy may be present.
- e) Liver, gingival, abdominal fat or rectal biopsy is done and gives positive Congo red test.

Differential diagnosis of hepatosplenomegaly in children :

- | | |
|----------------------------------|--|
| 1. Thalassemia major. | 7. Chronic malaria, chronic kala-azar. |
| 2. Acute leukaemias (ALL > AML). | 8. Wilson's disease. |
| 3. Indian childhood cirrhosis. | 9. Storage disorders like Gaucher's disease, glycogen storage disease. |
| 4. Acute viral hepatitis. | 10. Still's disease. |
| 5. Enteric fever. | 11. Congenital syphilis. |
| 6. Miliary tuberculosis. | |

N.B.: Always try to say that liver and spleen are 'palpable' below the costal margin. If you say they are enlarged, it means that the upper border of liver and splenic dullness are in normal position. If you detect the upper borders in normal position, you may say 'they are enlarged'.

There may be scar marks (rounded) present in the left and/or right hypochondrium in a chronic patient of hepatosplenomegaly due to application of counter-irritants by the quacks (used in remote villages) in a false attempt to treat the disease.

Case 34

CEREBELLAR DISORDER

Functions of cerebellum :

Cerebellum has ipsilateral control over body and,

1. Regulates the rate, rhythm, range and force of muscular contraction.
2. Facilitatory action on tone and reflex activity of the body.
3. Maintains posture and equilibrium.
4. Integrates the voluntary and automatic movements — 'coordination'.

Morphological and functional division of cerebellum :

1. Archicerebellum (oldest part)—Maintains locomotion and equilibrium (vestibular).
2. Palaeocerebellum (old part)—Maintains the muscle tone and posture (spinocerebellar).
3. Neocerebellum (youngest part)—Integration and coordination of fine muscular movements (corticocerebellar).

Bedside assessment of the cerebellar function :

A cerebellar hemispheric lesion will produce ipsilateral signs. Cerebellar function is assessed by :

1. **Titubation**—Nodding of head either in antero-posterior ('yes-yes') or side to side ('no-no') direction. Sometimes, there is 'head tilt' towards the side of lesion.
* Head-nodding may also be seen in aortic regurgitation (syphilitic), parkinsonism, mannerisms, anticonvulsants-induced and mental retardation.
2. **Speech (dysarthria)**—Dysarthria of staccato or scanning type. The speech is usually slow, slurred and irregular, and expressed with varying force. Often the patient scans the speech, i.e., he speaks syllable by syllable with undue emphasis on a particular syllable. Ask him to say 'artillery' or 'Motiur Rahaman': he will pronounce it as ar-til-ler-y or Mot-i-ur-Ra-ha-man.
3. **Hypotonia**—Elicit the tone of the muscle and it will be flaccid both at rest (flopy limbs) and on passive movements of the parts.
4. **Pendular knee jerk**—The patient will sit on a chair with legs hanging free side by side. Apply a sharp tap on the patellar tendon on each side, one after another. In health, contraction of quadriceps with extension of knee occurs. In case of cerebellar lesion, the knee jerks are very often diminished or pendular in nature. The first movement is followed by a series of diminishing oscillations (pendular) before the leg finally comes to rest. According to some clinicians, at least 3 to and fro movements in the leg are known as 'pendular'. If no response occurs, never forget to do the reinforcement test. Pendular knee jerk happens to be due to hypotonia.
5. **Intention tremor**—When tremor appears at the goal-point of an action, it is known as intention tremor. Ask the patient to hold a glass which is kept on the table or to hold your index finger which is kept away from him. The movements will be clumsy as he approaches the object. Finger-nose test also elicits intention tremor.
6. **Dysmetria**—It means inability to arrest the movements at desired point and is elicited by **'finger-nose test'**. The patient is asked to touch the tip of his nose with the tip of his index finger, first with eyes open and then with eyes closed (at the beginning of the test, the patient is directed to outstretch his arm and the procedure is demonstrated to him by the examiner). Instruct him to look straight ahead and not to focus on the index finger. Ask the patient to repeat the test as quickly as possible. This test can be modified by asking the patient to touch your index finger after touching his nose (finger-nose-finger test). First do the test on one side, then examine the other. The index finger of the patient may 'falls short' (hypometria) or 'overshoot' (hypermetria or past pointing) his nose. By this simple test one can assess the presence of :
 (i) Intention tremor,
 (ii) Coordination, and
 (iii) Side of lesion (by seeing the past pointing).
- 7 **Dyssynergia**—It is the inability to perform movements as a coordinated temporal sequences when the movements may be broken down into their component parts (decomposition of movements) producing small, jerky and clumsy movements (like the modern break-dance). The patient feels difficulty in performing complex movements.

8. **Dysidiadochokinesia** The patient can not execute rapid and repeated movements smoothly. He is asked to flex his elbows at right angle, and then pronate and supinate his forearm very rapidly 'as though screwing in a light bulb'. It can also be tested by asking the patient to supinate and pronate his right hand over the left palm very rapidly, and then the left hand over the right palm in the same way. In cerebellar lesion the movements are slow, clumsy, awkward, incomplete and often stops after few attempts (on the side of cerebellar lesion).
9. **Rebound phenomenon**—The limb overshoots beyond the normal range after sudden release of the resistance. Ask the patient to flex his elbow against resistance offered by you. As soon as you withdraw the resistance suddenly, the patient's hand tends to strike his face (because the antagonistic muscle like triceps can not contract promptly). This phenomenon is due to muscular hypotonia.
10. **Nystagmus** Horizontal, jerky nystagmus is present and the direction of nystagmus (fast component) is towards the side of lesion.
11. **Romberg's sign**—Actually it a test of sensory ataxia. The test is negative in cerebellar disease (see below).
12. **Gait**—
 - a) This is a 'reeling', 'drunken' or 'lurching' gait due to presence of truncal 'ataxia'. The patient walks on a broad base, the feet being placed widely apart and irregularly; at times, the head is tilted towards the side of lesion (try in those patients, who are able to walk).
 - b) Now the patient is asked to walk along a straight line (cracks already present in the floor or you draw a line there). The patient sways and often falls towards the side of lesion during walking.
 - c) Tandem gait' (heel to toe gait) —The patient is asked to walk along a straight line with the heel of one foot touching the toes of the one behind. The patient is then told to turn backwards quickly. It is very difficult for a patient with cerebellar lesion to walk steadily by tandem gait. This gait is a sensitive test of early ataxia (a test of vermis of cerebellum).

Presentation with cerebellar lesion :

Connections : Afferent fibres reach the cerebellum from spinal cord, vestibular system, basal ganglia and cerebrum. Cerebellum influences the LMN through its connections with the basal ganglia and cerebral cortex, via the thalamus. The common presentations are :

- a) Hemispherical lesion—**Paralysis** is not a feature of cerebellar disease rather the patient presents with ipsilateral hypotonia (weakness), incoordination (ataxia), and difficulty in walking or swaying while walking.
- b) Lesion at vermis—Produces truncal ataxia and the patient feels difficulty in sitting up or standing. Lesion at flocculonodular region produces vertigo, vomiting and gait ataxia, if they extend to the roof of 4th ventricle.

Features of lesion at vermis (midline lesion) :

In a midline cerebellar lesion there is little or no incoordination, no nystagmus and no hypotonia. But there are presence of :

- (i) Titubation (head-nodding),
- (ii) Truncal ataxia (ataxia of the body),
- (iii) Positive 'heel to toe' gait (tandem gait), and
- (iv) Difficulty in sitting, standing and unassisted walking (swaying and unsteadiness).

} 3 T

What is sensory ataxia ?

For the production of a purposeful coordinated movement the cortex, the cerebellum, the reflex arc, impulses from eyes, labyrinth and cervical spine are required to function together. If such coordination is imperfect, motor performance becomes difficult. **Clumsiness of the movement {clinically manifested by unsteady gait} is known as ataxia provided, there is no motor and sensory deficit** Ataxia is mainly of four types :

- a) Cerebellar — Vascular lesion, cerebellar tumour, cerebellitis, cerebellar degeneration, hereditary ataxias, alcohol or phenytoin-induced.
- b) Sensory — Peripheral neuropathy, tabes dorsalis.
- c) Labyrinthine — Acute labyrinthitis, streptomycin-induced, Meniere's disease.
- d) Central — Affection of vestibular nucleus as a result of vascular lesion in the medulla.
- e) Miscellaneous — Severe muscular weakness, hypotonia, alcoholism.

Sensory ataxia results from defective proprioception (due to posterior column involvement) and can sometimes be alleviated with the help of vision. Thus sensory ataxia is increased when the eyes are closed, in contrast to cerebellar ataxia which is not affected by vision. Tabes dorsalis is a classical example of sensory ataxia.

Sensory ataxia is usually due to affection of :

- a) Peripheral sensory nerves —Peripheral neuropathy (e.g., leprosy, diabetes mellitus).
- b) Posterior nerve roots —Tabes dorsalis, disc prolapse.
- c) Posterior column —Multiple sclerosis, syringomyelia.
- d) Post-central convolution —Diseases of parietal lobe (vascular, SOL).

* **[Apraxia]** Inability to perform certain purposive well-organized voluntary movement in the absence of motor weakness, sensory loss, cerebellar or extrapyramidal impairment, or ataxia. It develops in frontal and parietal lobe lesions. Ask the patient to light a cigarette with a matchstick, or copy a simple diagram. It seems, in apraxia, the person has forgotten to perform the motor act.

** **Agnosia**—Inability to recognise the nature of sensory input in spite of intact sensory organ, e.g., visual agnosia, tactile agnosia and auditory agnosia.)

What is Romberg's sign ?

It is important to remember that **it is a sign of sensory ataxia** (i.e., a test of posterior column function) and not a test for cerebellar function.

The patient is asked to stand with his feet close together (convergent heels with divergent toes). If he can do this, he is then asked to close his eyes. Romberg's sign is said to be present or positive when the patient begins to sway or about to fall as soon as he closes his eyes. **The cardinal feature of this sign is that "the patient is more unsteady standing with his eyes closed than when the eyes are kept open** . A patient with cerebellar ataxia or labyrinthine lesion shows little or no increase in instability after closing the eyes (though the patient may show some unsteadiness at the beginning of the test with open eyes). False positive Romberg's sign is observed in hysteria (hips sway more in comparison to ankles).

Prerequisites :

1. Patient will stand bare-footed,
2. Feet placed close to each other.
3. The examiner will stand very close to the patient and assure the patient of his safety.

Table 23 : Differentiation between cerebellar and sensory ataxia

Features	Cerebellar ataxia	Sensory ataxia
1. Power of muscles	Normal	May be diminished (rarely)
2. Jerk (deep reflexes)	Pendular	Lost
3. Cerebellar signs	Present	Absent
4. Proprioceptive sensations	Present	Lost
5. Charcot joint and trophic changes	Absent	Present
6. Romberg's sign	Negative	Positive
7. Gait	Reeling gait	Stamping gait
8. Examples	Friedreich's ataxia. multiple sclerosis	Tabes dorsalis, peripheral neuropathy

Features of posterior column lesion :

1. Motor functions :
 - a) Nutrition — Mild wasting.
 - b) Tone — Diminished.
 - c) Power — Normal (i.e., preserved).
 - d) Involuntary movements — Absent; rarely pseudo-athetotic movements are seen.
 - e) Coordination — Usually normal.
2. Sensory functions :
 - a) Touch (crude), pain and temperature — Unaffected.
 - b) Deep sensations (joint, position, vibration sense) with fine touch — Lost.
 - c) Girdle-like sensation at the level of compression—Present.

- d) Cortical sensations—Lost.
- e) Lhermitte's sign — momentary electric shock-like sensation on the back (the spine and into the legs) evoked by neck flexion, and is also known as “Barber's chair sign (causes of Lhermitte's sign are multiple sclerosis, cervical spondylosis, subacute combined degeneration, cervical cord tumour and syringomyelia).
- 3. Reflexes :
 - a) Superficial — Normal.
 - b) Deep reflexes or jerks — Diminished or may remain normal.
 - c) Plantar response — Flexor.
 - d) Bladder and bowel — Normal.
- 4. Trophic changes — May be present.
- 5. Romberg's sign - present. Sensory ataxia and stamping gait.
- * Loss of proprioception = ‘Rombergism’.

Diseases involving the posterior column :

Tabes dorsalis, subacute combined degeneration, cervical spondylosis, compressive myelopathy (posteriorly located), acute transverse myelitis etc.

How to test for coordination :

(A) In the upper limbs :

1. Watch the patient while dressing or undressing, picking up pins from the table, combing etc.
2. Threading a needle.
3. Finger-nose test.
4. Dysidiadochokinesia.

(B) In the lower limbs :

1. To make a circle in the air with the foot while in lying down position.
2. Walking along a straight line and tandem gait.
3. Heel-knee test.
4. Romberg's sign.

* Remember, muscular weakness may be accompanied by clumsiness of movement which may pose difficulty in interpretation of coordination.

What is heel-knee (heel-shin) test ?

The patient lies in bed. Ask him to raise one leg high in the air and then place the heel of the flexed leg on the opposite knee and to then slide the heel down the anterior surface of leg (shin) towards the ankle upto the great toe. Before doing the test, demonstrate it clearly to the patient. Now ask the patient to do the test on the other side. In health, the movement of the heel is smooth and uniform throughout. In cerebellar disorder, the test becomes irregular, and there are errors in the direction and speed of movement.



Common cerebellar disorders :

1. Cerebellar neoplasm (medulloblastoma, haemangioblastoma).
2. Multiple sclerosis.
3. Viral cerebellitis (e.g., post chicken-pox cerebellitis).
4. Cerebellar abscess.
5. Friedreich's ataxia.
6. Other spinocerebellar degeneration (signs of pyramidal tract lesion along with cerebellar signs).
7. Vascular—Thrombosis of the posterior inferior cerebellar artery, cerebellar haemorrhage.
8. Cerebello-pontine angle tumours.
9. Paraneoplastic syndrome.

Acquired causes of cerebellar degeneration :

1. Myxoedema.
2. Phenytoin toxicity.
3. Alcoholic degeneration (chronic alcoholism).
4. Neoplasm anywhere in the body e.g., bronchogenic carcinoma (paraneoplastic syndrome).
5. Infection (HIV, abscess).

Case 35

BULBAR PALSY

What is bulbar palsy ?

Paralysis or loss of function of the muscles supplied by the cranial nerves arising from the bulb (old name for medulla oblongata) i.e., it is a palato-labio-glosso-pharyngo-laryngeal paralysis. Truly speaking, the motor nuclei of the IXth, Xth, XIth and XIIth cranial nerves are affected in bulbar palsy. So it is a **LMN type of paralysis**.

What are the major symptoms ?

1. Dysarthria (slurring of speech),
2. Dysphonia or nasal intonation,
3. Dysphagia, and
4. Nasal regurgitation.

* Choking is another common symptom.

What is pseudobulbar palsy ?

It is the bulbar paralysis due to involvement of bilateral pyramidal tracts in the brain stem or above. So in this **UMN type of lesion**, the voluntary control of the muscles supplied by the IXth, Xth, XIth and XIIth cranial nerves will be impaired.

Clinical features of IXth, Xth, XIth and XIIth cranial nerves palsy :

THE RULE OF THREE' :

(A) Glossopharyngeal nerve—

1. Loss of sensation over the mucous membrane of pharynx.
2. Loss of taste sensation in the posterior 1/3rd of the tongue.
3. Do the 'gag reflex' (the afferent is IXth and the efferent is Xth cranial nerve; centre for the reflex lies in medulla) and examine for taste sensation in the posterior 1/3rd of the tongue. **Gag reflex**—Depress the tongue with a spatula. Touch the posterior wall of pharynx or tonsil by a piece of cotton wrapped in a broom-stick and note its reflex contraction. Normally there is bilaterally symmetrical contraction of posterior pharyngeal wall and fauces of mouth. Test each side separately, and ask for appreciation of touch sensation. This reflex is lost in bulbar palsy.

(B) Vagus nerve—

- a) Soft palate—
 1. Ask the patient for nasal intonation (twang) and hoarseness of voice.
 2. Ask the patient for nasal regurgitation of liquids.
 3. Ask the patient to say 'aah' ! and observe whether both the sides of the palate arch upwards or not.
- b) Pharynx—
 1. Dysphagia—Maximum to liquid and minimum to semisolid food, in contrast to mechanical dysphagia where dysphagia is maximum with solid food (liquid food needs immediate muscular contraction to swallow and this is why neurogenic dysphagia occurs earlier with liquid food).
Moreover, pooling of secretions in the posterior pharynx is noted with drooling of saliva; there may be sensation of choking.
 2. Nocturnal cough—During sleep, saliva goes into the respiratory tract and may precipitate death due to pneumonia.
 3. Test the 'gag reflex'.
- c) Larynx—
 1. Hoarseness of voice (adduction of vocal cord is not possible); bovine cough.
 2. Inspiratory dyspnoea (abduction of vocal cord is not possible).
 3. Examine the vocal cord by indirect laryngoscopy.

(C) Spinal accessory nerve—

1. In bilateral paralysis of sternomastoid muscle, head tends to fall back.
 2. Test the power of sternomastoids from front.
 3. Test the power of trapezius from behind.
- Read the section on 'Myotonia' for proper testing of these muscles.

(D) Hypoglossal nerve—

1. Ask the patient to protrude the tongue — Look for any deviation (deviated to the side of paralysis due to unopposed action of genioglossus of healthy side). The patient may not be able to protrude the tongue much beyond the teeth in bilateral XIIth nerve palsy.
2. Ask the patient to curl his tongue up and down, and sideways. Test the power of the tongue muscles (ask the patient to bulge his cheeks with the tongue against resistance offered by the examiner from outside).
3. While the tongue is kept within the oral cavity, observe for wasting and fasciculation.

Causes of pseudobulbar and bulbar palsy :**(A) Pseudobulbar palsy :**

1. Diffuse cerebral ischaemia (multi-infarct dementia) — Lacunar infarction.
2. Diffuse atherosclerosis (cerebral atrophy).
3. Chronic motor neurone disease (MND).
4. Sometimes in multiple sclerosis (a late event).
5. Cerebral vasculitis.
6. Following severe brain injury.

(B) Bulbar palsy :**a) Acute —**

1. Poliomyelitis (bulbar variety).
2. Diphtheria.
3. G. B. syndrome.
4. Rabies.
5. Organophosphorus poisoning.
6. Encephalitis.
7. Snake bite (Elapidae),
8. Botulism.
9. Myasthenic crisis.
10. Medullary infarction.

b) Chronic —

1. Chronic motor neurone disease (MND).
2. Syringobulbia.
3. Neoplasm in the medulla or pontine glioma.
4. Basal meningitis.
5. Neurosyphilis.
6. Fracture of the posterior cranial fossa.
7. Multiple metastasis involving lower cranial nerves.

* Intermittent bulbar palsy—may be seen in myasthenia gravis and multiple sclerosis.

** MND can develop into both pseudobulbar (UMN) and bulbar (LMN) palsy.

Other features to note in bulbar palsy :

The features developing due to involvement of vital centres in the medulla should be carefully monitored (mainly in acute variety).

1. Cardiac centre—Tachycardia, different arrhythmias.
2. Respiratory centre—Tachypnoea, ataxic breathing, respiratory failure.
3. Vasomotor centre—Hypotension or hypertension.

There may be damage to fibre tracts like pyramidal tract (crossed hemiplegia or decerebrate rigidity), sympathetic trunk (ipsilateral Horner's syndrome), spinothalamic tract (loss of touch, pain, temperature sensation on the opposite side), medial lemniscus (ipsilateral loss of proprioception), vestibulospinal tracts (vertigo), descending tract and nucleus of the Vth cranial nerve (ipsilateral touch, pain and loss of temperature sensation in the face), and nystagmus.

Reflexes in pseudobulbar and bulbar palsy :

1. Superficial reflex i.e., plantar response is always extensor in pseudobulbar palsy and usually flexor in bulbar palsy (may be extensor, depending on the aetiology e.g., in motor neurone disease).
2. Deep reflex i.e., Jaw jerk is brisk in pseudobulbar palsy and absent in bulbar palsy (a very important sign for differentiation at bedside).

Table 24 : Differentiation between pseudobulbar and bulbar palsy

Features	Pseudobulbar palsy	Bulbar palsy
1. Age	1. Usually above 60 years	1. Depends on the aetiology (usually middle aged)
2. Site of lesion	2. Cortex and subcortex	2. Medulla
3. Type of lesion	3. UMN type	3. LMN type
4. Onset	4. Always gradual	4. Acute or gradual
5. Memory loss and emotional instability	5. Present	5. Absent
6. Blood pressure	6. May be high	6. Often normal
-7. Facies	7. Masked facies	7. Nothing suggestive
8: Dysarthria	8. Spastic	8. Flaccid
9. Tongue	9. 'Spastic' and firm with pointed tip, there is neither wasting nor any fasciculation seen; small tongue	9. 'Flaccid', wasted, flabby with rounded tip, movements grossly diminished (may occupy one corner of the mouth); fasciculation +
10. Jaw jerk	10. Brisk	10 Absent
11. Plantar response	11. Extensor	11 Flexor or extensor (depends on aetiology)
12. Bulbar muscle involvement	12. Milder degree	12. Moderate to severe involvement

Disease with combined bulbar and pseudobulbar palsy-features :

Chronic motor neurone disease (amyotrophic lateral sclerosis) may have combined palsy and is characterised by wasted tongue with brisk jaw jerk.

Management of respiratory paralysis with or without bulbar palsy :

A patient suffering from respiratory paralysis should be treated by artificial respiration which may be required for weeks or months, or indefinitely :

1. Intermittent positive pressure ventilation through a nasal or oral endotracheal tube (when needed for 2-3 days) or through a cuffed tube inserted via a tracheostomy, preferably in a respiratory care unit (RCU) with regular monitoring of blood gases to avoid over- or under-ventilation. Cuffed tube prevents the passage of pharyngeal secretions into the lungs.
2. Appropriate antibiotics to prevent pneumonia.
3. Care of the skin, bladder and bowel.

* Treatment of bulbar palsy requires :

4. Nasogastric suction by mechanical sucker.
5. Postural drainage — Foot end of the bed should be raised and the patient is turned in the semi-prone position.
6. Proper fluid balance.
7. Chest physiotherapy.

Causes of nasal regurgitation of fluid :

1. Paralysis of vagus nerve.
2. Bulbar and pseudobulbar palsy.
3. Polymyositis or dermatomyositis.
4. Myopathy.
5. Cleft palate.
6. Perforation or destruction of palate (syphilis, tuberculosis, leprosy).
8. Malignant disease of nasopharynx.

* Common causes of bulbar palsy : G. B. syndrome, MND, snake bite, diphtheria and poliomyelitis.

** The gag and palatal reflexes are preserved in pseudobulbar palsy.

N.B. : Bach of the long cases e.g., valvular heart diseases, pleural diseases, lymphoma, thalassaemia etc may be treated as short cases.

CHAPTER III : SPOT CASES

Case 36

ANAEMIA

Definition :

It is defined as the qualitative or quantitative diminution of RBC and / or haemoglobin concentration in relation to standard age and sex, and is clinically manifested by pallor. It is not a disease but denotes manifestation of some disease, and thus a cause must be searched for.

Where will you look for anaemia ?

Sites :

1. Lower palpebral conjunctiva (retract the lower eyelids downward and ask the patient to look upwards — both eyes at a time).
2. Tongue, specially the tip and the dorsum.
3. Mucous membrane of palate.
4. Nail-beds (press the pulp to see the redness of nail-bed).
5. Palms, soles and general skin surfaces.

* Nail-beds are the windows of the cutaneous capillary network.

Clinical types of anaemia and its bedside assessment :

Anaemia is clinically classified as mild, moderate and severe types. It is totally a clinical assessment (highly subjective sign) and may not correspond with laboratory findings. Clinically we see the paleness of the skin/mucous membrane, or pallor. The colour of the tongue as well as the conjunctiva are more reliable than other sites in this respect (in adults). In children, palms and soles are to be specially looked for.

Clinical classification :

Mild anaemia — 60-80% of Hb (9-12 g/dl).

Moderate anaemia — 40-60% of Hb (6-9 g/dl).

Severe anaemia — < 40% of Hb (< 6 g/dl).

The normal haemoglobin concentrations in case of males and females are.

Males — 14.6 to 15.5 g/dl, and females — 13.3 to 14.6 g/dl.

* Clinically, 14.5 g/dl may be taken as 100%.

Causes of 'pallor without anaemia' :

See the section on 'Myxoedema'.

* **Pallor** (paleness) is the waxy appearance of skin and mucous membrane. It depends on thickness and quality of skin, and quality and amount of blood in the capillaries. Thus, pallor and anaemia are not interchangeable terms. There are many causes of pallor, and anaemia is commonest of them. Pallor of mucous membrane is commonly due to anaemia; very often generalised pallor is attributed to severe anaemia. It is difficult to detect pallor in deeply pigmented individuals.

** Anaemia is a pathological condition while pallor is a clinical entity. A person without losing a drop of blood may become deadly pale (shock and collapse), and similarly a person looking severely pale may not be grossly anaemic (Sheehan's syndrome).

*** Facial pallor is commonly seen in shock and low cardiac outputs state.

general survey, one may write pallor (probably a better terminology) instead of anaemia.

Examination of the hands in anaemia :

1. Colour nail-beds and palms give an indication of presence of anaemia.
2. Koilonychia gives a clue to the type of anaemia, such as iron deficiency anaemia.
3. In severe anaemia, there may be capillary pulsation.
4. Presence of splinter haemorrhage or Osier's node points towards SBE.

5. Anaemia with finger deformities may indicate rheumatoid arthritis.
 6. Colour of palmar creases gives a clue to the degree of anaemia. When they are as pale as the surrounding skin, the patient usually has haemoglobin level < 7 g/dl.
- * In ophthalmoscopy if the retina seems very pale, the haemoglobin concentration is usually < 4 g/dl.
- ** Jaundice associated with anaemia points towards haemolytic anaemia.

Haemoglobin concentration is 81%-99% : is it anaemia ?

Yes, it is anaemia but the pallor may not be clinically evident (so this group is not included in the clinical classification of anaemia).

Symptoms referable to anaemia :

Weakness, fatigue, lassitude, light-headedness, giddiness, fainting or syncope, anorexia, palpitation, breathlessness, anginal pain, insomnia, tinnitus, lack of concentration, intermittent claudication, tingling sensation in the extremities and menstrual irregularities are common symptoms.

Importance of 'history taking' in a patient of anaemia :

1. Present history—commonly complaining of easy fatigability, shortness of breath and decreased effort intolerance.
2. Past history—H/O bleeding (e.g., menorrhagia or haemorrhoids), H/O blood transfusion in the past or diarrhoea (tropical sprue).
3. Personal history—alcohol consumption, walking bare-footed (e.g., hookworm infestation).
4. Family history—similar type of illness in the family suggests genetic inheritance, e.g., thalassaemia, HbE disease, haemophilia.
5. Dietary history—should be taken to diagnose nutritional anaemia (e.g., true vegetarians may suffer from vitamin B₁₂ deficiency anaemia); less intake due to anorexia, dysphagia or poverty.
6. Treatment history—Bone marrow suppressant drugs, NSAID-intake or H/O radiation.
7. Occupational history—H/O exposure to chemical solvents, lead, benzene, insecticides should be taken; a cultivator working bare-footed may suffer from hookworm infestation.
8. Geographic background and ethnic origin—Important to diagnose thalassaemia, sickle cell anaemia, G₆PD deficiency.

Important history to evaluate anaemia :

H/O chronic blood loss (bleeding peptic ulcer, haemorrhoids, menorrhagia), malabsorption (diarrhoea/steatorrhoea), nutritional intake, worm infestations, exposure to drugs/chemicals/radiation, bleeding tendencies, bowel habits, anorexia and loss of weight.

Common causes of severe anaemia in your hospital :

- | | |
|-----------------------------------|--|
| 1. Nutritional. | 6. Acute leukaemias. |
| 2. Hookworm infestation. | 7. Thalassaemia. |
| 3. Haematemesis and melaena. | 8. Uraemia. |
| 4. Chronic bleeding haemorrhoids. | 9. Menorrhagia. |
| 5. Aplastic anaemia. | 10. Carcinoma of stomach, lung, colon. |

Importance of stool examination in anaemia ;

1. Presence of occult blood.
2. To diagnose hookworm ova.

Classify anaemia (working classification) :

- (A) Based on morphology : normocytic, microcytic, macrocytic.
- (B) Based on aetiology :

- a) Blood loss, which may be acute or chronic.
- b) Inadequate production of RBC.
- c) Excessive destruction of RBC.

* It seems anaemia may result from nutritional deficiency, haemorrhage, haemolysis or hypoplasia of the bone marrow.

Cardiovascular (CVS) features in severe anaemia :

1. Tachycardia.

2. Capillary pulsation.
3. Water-hammer pulse.
4. Cervical venous hum.
5. Cardiomegaly (cardiac dilatation) and later signs of heart failure.
6. Hyperdynamic apex beat.
7. Mitral systolic murmur due to functional MI (ring dilatation).
8. Haemic murmur over the pulmonary area.
9. Rarely mid-diastolic (non-rumbling) murmur in mitral area may be auscultated (due to relative stenosis at mitral valve) secondary to Increased blood flow.

* No. 1, 3, 5 and 8 are most important.

Features in other systems in severe anaemia :

- | | |
|---------------------------|--|
| 1. General survey | a) Pallor b) Dyspnoea c) Ankle oedema or anasarca. |
| 2. CVS | Already described above. |
| 3. G.I. tract | Hepatosplenomegaly may be present. |
| 4. Respiratory system | Basal crepitations. |
| 5. Nervous system | Features of polyneuropathy; sometimes, there is papilloedema |
| 6. Lymphoreticular system | There may be presence of sternal tenderness. |

Ankle oedema in anaemia :

1. Severe anaemia per se (renal retention of salt and water).
2. Congestive cardiac failure may be associated with.
3. Associated renal or hepatic disorders, hypoalbuminaemia, myxoedema.

Common causes of anaemia in this part of world :

1. Nutritional anaemia (so, dietary history is important).
2. Hookworm infestation.
3. Chronic malaria, chronic kala-azar, tuberculosis.
4. Thalassaemia.
5. Tropical sprue.

* Transfusion is usually required when Hb level goes below 7 g/dl. One unit (2 units in India) of packed red cells elevates the haemoglobin by approximately 1 g/dl.

** Globally, 'chronic blood loss' is probably the commonest cause of anaemia.

Common causes of iron deficiency anaemia :

- | | |
|------------------------------|----------------------------|
| 1. Nutritional deficiency. | 5. Hookworm infestation. |
| 2. Bleeding peptic ulcer. | 6. Malabsorption syndrome. |
| 3. Gastric erosion by NSAID. | 7. Menorrhagia. |
| 4. Bleeding haemorrhoids. | 8. Pregnancy, lactation. |

* This is why stool is examined for ova/parasite/cyst/occult blood, upper and lower G.I. endoscopy are performed with barium study, and gynaecological evaluation are done for 'chronic blood loss'.

Clinical diagnosis of iron deficiency anaemia :

1. H/O 'pica' (eating of strange non-nutrient items such as clay—geophagia, ice—pagophagia, cornstarch—amylphagia, or coal) or chronic blood loss from G. I. tract.
2. Pallor (pearly white sclera).
3. **Glossitis** (bald or depapillated tongue)
4. *Angular stomatitis*, cheilosis.
5. Brittle finger nails, flat nails (platynychia) or even *koilonychia*.
6. Brittle hairs.
7. Dysphagia (Plummer-Vinson syndrome or Paterson-Kelly syndrome) as a result of post-cricoid web which can be demonstrated endoscopically or by barium swallow.
8. Mild splenomegaly, rarely.

* 'Pica' (i.e., perverted appetite) is also observed in pregnancy and some psychiatric disorder.



Enucleation of the left eye



Enophthalmos (right) – phthisis bulbi of the right eye resulting from smallpox in childhood



← Massive **cervical adenopathy** producing bull-neck from lymphoma; infiltration of chest and anterior abdominal skin resulted in 'lymphoma cutis'



Cavernous sinus thrombosis in the left side resulting in proptosis, ptosis and ophthalmoplegia



A moribund patient showing **proptosis** due to involvement of the right eye from lymphoma (NHL); cervical adenopathy is also noted

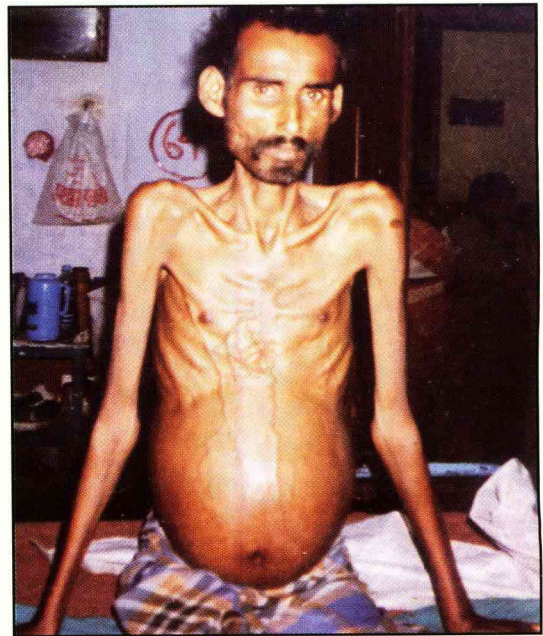


SVC syndrome. Note the prominent external jugular veins, and veins over the anterior chest wall. The face is puffy, and chest and upper limbs are swollen. The patient had moderate degree of respiratory distress



IVC obstruction. Prominent collateral veins are seen over the anterior abdominal walls and flanks. There is absence of ascites

→
Portal hypertension. Prominent and tortuous veins over the abdomen as well as ascites are present. The patient is grossly emaciated – a classical case of cirrhosis of liver



Portal venous obstruction showing prominent collaterals. Venous hum is audible over the visible veins



Engorgement of both external and internal **jugular veins** in a patient with congestive cardiac failure

Causes of glossitis and angular stomatitis :**(A) Glossitis (inflammation with red and sore tongue) :**

- a) Riboflavin deficiency—Magenta-coloured tongue.
- b) Iron deficiency anaemia.
- c) Nicotinic acid or vitamin B₁₂ deficiency—The tongue is red, swollen, painful and is known as 'raw-beefy tongue'.
- d) Folic acid deficiency.
- e) Bald tongue (complete atrophy of papillae)—Found in severe iron deficiency anaemia, tropical sprue, pernicious anaemia, pellagra and syphilis.
- f) Median rhomboid glossitis— A lozenge-shaped denuded area in the middle of the tongue posteriorly. It is a congenital abnormality and as it feels nodular, it should be differentiated from carcinoma of the tongue.
- g) Miscellaneous—cirrhosis of liver, tuberculosis.

(B) Angular stomatitis (inflammation of the skin at the angle or corner of the mouth) :

- a) Excessive use of tobacco, alcohol or betel-leaf.
- b) Improperly-fitted denture.
- c) Iron deficiency anaemia.
- d) Riboflavin, nicotinic acid, folic acid and pyridoxine deficiency.
- e) Starvation or malnutrition.
- f) Herpes labialis, candidiasis or streptococcal infection at the angle of mouth.
- g) 'Perleche', in children, are infective in origin.

Causes of dimorphic anaemia :

Dimorphic anaemia is macrocytic hypochromic in type (mixture of iron deficiency and megaloblastic anaemia). Common causes are,

1. Pregnancy.
2. Hookworm anaemia with nutritional deficiency.

Causes of haemolytic anaemia i

See the sections on 'Jaundice' and 'Thalassaemia'.

How do you diagnose hookworm infestation ?

1. Occupation e.g., patients working in tea-garden, coal mines or a farmer who walks bare-footed.
2. Presence of ground itch at the site of entry in the skin.
3. Perversion of taste sensation with 'pica' (often they eat earth, mud or lime).
4. Blotting paper like tongue (very pale tongue) with puffy face and swelling of lower eyelids. Koilonychia and pedal oedema are frequently observed. There is lustreless hair with protuberant abdomen. Other features of iron deficiency anaemia may be present.
5. Pain in the epigastrium mimicking chronic duodenal ulcer.
6. Steatorrhoea or fatty diarrhoea.
7. Passage of small worms in stool.

Causes of anaemia due to chronic systemic diseases :**(A) Anaemia of chronic inflammation :**

- a) Infection—SBE, lung abscess, tuberculosis, osteomyelitis, pyelonephritis.
- b) Collagen vascular disorders—SLE.
- c) Malignancy.
- d) Rheumatoid arthritis.

(B) Uraemia.**(C) Endocrine disorders—Myxoedema, Addison's disease, panhypopituitarism.****(D) Liver disease—Specially in alcoholics.**

The whole spectrum is associated with normocytic normochromic anaemia. Group (A) may have microcytic anaemia. Group (B) may have Burr cells, and Group (C) and (D) may show macrocytic RBCs. The total group will have low serum iron, low TIBC, and normal or elevated serum ferritin level.

What is 'leucoerythroblastic' blood picture ?

When normoblasts and myelocytes both are seen in the peripheral blood of patients suffering from myelophthisic anaemia (infiltration of bone marrow by tumour or granuloma), M_6 variety of AML, myelofibrosis or myelosclerosis, it is said that leucoerythroblastic blood picture is present.

Outline of investigations in anaemia :

The investigations are directed to see the type and aetiology of anaemia :

- (A) The type of anaemia is ascertained by peripheral blood smear, blood indices and serum values
 - a) Peripheral blood smear—RBC morphology, reticulocytes, malaria parasite.
 - b) Blood indices—MCV, MCH, MCHC.
 - c) Serum values—serum iron concentration, TIBC, serum ferritin.
- (B) The cause of anaemia is determined by meticulous history taking, clinical examination, haematological tests (described above), examination of stool/urine, chest X-ray, special haematological tests (Coombs' test, osmotic fragility test, Hb-electrophoresis etc), and in selective cases by bone marrow study.

How do you examine a patient for polycythemia ('ruddy cyanosis') ?

Polycythemia is the increase in red blood cells above normal.

It is examined in the same way as done for the clinical detection of anaemia i.e., look for the lower palpebral conjunctiva, tongue, palate, nail-beds, palms and soles, and general skin surface. The colour of the mucous membrane turns dusky-red. Patients of polycythemia usually have facial plethora, suffused conjunctiva, increased redness of lower palpebral conjunctiva and palmar erythema.

Common causes of polycythemia in tertiary care hospitals :

1. COPD.
2. Cyanotic congenital heart diseases e.g., Fallot's tetralogy.
3. Right-to-left shunt in the heart.
4. Polycythemia vera.

* High altitude and severe dehydration (relative polycythemia) are also common causes of polycythemia. Thrombosis, peptic ulceration (often with bleeding) and hyperuricaemia are noted complications of polycythemia vera.

Anaemia and polycythemia in a single patient—is it possible ?

Sometimes it is seen in Fallot's tetralogy where anaemia (iron deficiency) is not clinically evident in the presence of polycythemia. If Hb% is < 18 g/dl, PCV is < 55%, and MCHC is < 28 g/dl, one should be suspicious about anaemia in the presence of polycythemia in a case of Fallot's tetralogy.

Normal reticulocyte count, reticulocyte index and red cell mass (volume) :

- | | |
|-------------------------|---|
| (A) Reticulocyte count— | 0.2-2% of RBCs (increased in haemolytic anaemia). |
| (B) Reticulocyte index— | $\text{Reticulocyte \%} \times \frac{\text{Patient's PCV}}{\text{Normal PCV}}$ |
| (C) Red cell mass— | <ol style="list-style-type: none"> (i) Male - 30 ml/kg of body weight. (ii) Female - 25 ml/kg of body weight. (increased in polycythemia vera). |

Conclusion :

Always examine both the lower palpebral conjunctiva because the presence of conjunctivitis in one eye may interfere with the interpretation of anaemia if anyone examines the infected eye only. While examining the tongue for anaemia, very often magenta-coloured tongue (riboflavin deficiency) misguides a clinician. By retracting the lower eyelids downwards, one can diagnose two other conditions e.g., polycythemia (increased redness of conjunctiva) and scleroderma (increased tightness of the skin during retraction). Do not examine the eyes for anaemia if it is rubbed or washed just before testing.

Earliest sign of acute blood loss is tachycardia and postural hypotension, and not anaemia because haemodilution (resulting anaemia) is completed in 24-36 hours.

Case 37

CYANOSIS

What is cyanosis ?

It is defined as bluish discolouration of the skin and mucous membrane due to presence of increased amount of reduced haemoglobin (> 5 g/dl), or of haemoglobin derivatives in the capillary blood. Absolute concentration of reduced haemoglobin above 5 g/dl in the capillaries makes the blood dark and gives the tissue a bluish hue ('kyaan' means dark blue colour and 'osis' means condition). According to few clinicians, > 4 g/dl of reduced (desaturated) haemoglobin in the capillary blood produces cyanosis (recent view).

Cyanosis is a grave sign and needs immediate evaluation as well as intervention.

Types of cyanosis :

It is principally of two types : Central and peripheral cyanosis.

* Other clinical types of cyanosis are : enterogenous, mixed and differential cyanosis.

Peripheral cyanosis and sites to be examined :

In peripheral cyanosis, the arterial blood is normally saturated (**arterial PaO_2 is normal**) but there is oxygen unsaturation at the venous end of capillary. Peripheral cyanosis results from excessive extraction of oxygen from the normally saturated arterial blood. The different mechanisms play in its production are,

- a) Reduced cardiac output.
- b) Peripheral vasoconstriction.
- c) Slow speed of circulation in the extremities.

Sites to be looked for (in good natural light)—

1. Tip of the nose.
2. Ear lobules.
3. Outer aspect of lips, chin and cheek.
4. Tip of fingers and toes.
5. Nail-bed of fingers and toes.
6. Palms and soles.

Tongue remains unaffected. Mechanism : stagnant as well as over-utilisation hypoxia.

Causes of peripheral cyanosis :

1. Exposure to cold air or cold water (possibly the most common cause).
2. Congestive cardiac failure (reduced cardiac output).
3. Frost bite.
4. Raynaud's phenomenon (read the section on 'Scleroderma')—only seen in fingers.
5. Shock or peripheral circulatory failure due to any cause.
6. Venous obstruction—produces local cyanosis (e.g., SVC syndrome).
7. Hyperviscosity syndrome (e.g., multiple myeloma, polycythemia, macroglobulinaemia).
8. Arterial obstruction e.g., peripheral vascular disease (atherosclerosis, Buerger's disease, atheroembolism).
9. Cryoglobulinaemia (abnormal globulin which forms gel at low temperature e.g., lymphoma).
10. Mitral stenosis (lips, tip of the nose and cheeks may be cyanosed in mitral facies).
11. Septicaemia (gram negative organisms commonly).

Central cyanosis and sites to be examined :

There is decreased arterial oxygen saturation as a result of marked reduction in oxygen tension in the arterial blood (**arterial PaO_2 is reduced**) either due to imperfect oxygenation of blood in lung or admixture of venous and arterial blood. This is usually detected when the oxygen saturation of arterial blood goes below 80-85%.

Sites to be looked for (in good natural light)—

1. Tongue (mainly the margins as well as the undersurface).
2. Inner aspect of lips.

3. Mucous membrane of gum, palate, cheeks.
4. Lower palpebral conjunctiva.
5. **Plus**, the sites mentioned in the peripheral cyanosis (one must examine these sites)—as same deoxygenated blood circulates everywhere in the body i.e., patients with central cyanosis will also be cyanosed peripherally.

* Mechanism : hypoxic hypoxia.

** In dark-skinned persons, examination of oral mucous membrane and conjunctiva are more helpful.

Causes of central cyanosis :

1. Cyanotic congenital heart diseases e.g., Fallot's tetralogy, transposition of great vessels.
2. Acute pulmonary oedema (due to left-sided heart failure)—most common 'cardiac cause.
3. Eisenmenger's syndrome (ASD, VSD or PDA with reversal of shunt due to development of pulmonary hypertension).
4. Acute severe asthma.
5. COPD, cor pulmonale, respiratory failure; respiratory depression (e.g., opium poisoning).
6. Lobar pneumonia.
7. Fibrosing alveolitis.
8. Tension pneumothorax.
9. Acute laryngeal oedema.
10. Acute pulmonary thromboembolism.
11. Pulmonary arteriovenous fistula (congenital, or acquired due to cirrhosis of liver).

* Never forget, high altitude (due to low partial pressure of O_2) as a cause of central cyanosis.

** In arterial blood, 95% of Hb is saturated with O_2 (i.e., only 5% reduced) whereas in venous blood the O_2 saturation is 70% (i.e., 30% reduced). Hence normally a mean of the two (5% to 30%, i.e., 0.75 g/dl to 5 g/dl reduced Hb) is present in the capillaries, and skin and mucous membrane remain pink. If the level crosses > 5 g/dl, the skin becomes blue or dusky.

What is 'enterogenous cyanosis' or pigment cyanosis ?

When cyanosis is produced due to the presence of excessive sulphaemoglobin (> 0.5 g/dl) or methaemoglobin (> 1.5 g/dl) in the blood, it is known as enterogenous cyanosis. Few clinicians include it within central cyanosis. The causes are :

1. Hereditary haemoglobin M disease.
2. Poisoning by aniline dyes (often added in sweets).
3. Drugs like nitrates and nitrites (nitroglycerin, sodium nitroprusside, amyl nitrite), phenacetin, sulphonamides, dapsone; nitrite may be present in well-water.
4. Carboxyhaemoglobinaemia (a cherry-red flush common in smokers; not true cyanosis).

* Diagnosis is confirmed by spectroscopic examination of the blood.

Differential diagnosis of bluish discolouration of body :

1. Cyanosis.
2. Carbon monoxide poisoning—Actually it is a cherry-red flush due to carboxyhaemoglobin.
3. Argyria—Deposition of silver salts in the skin due to silver poisoning. The skin does not blanch on pressure (cyanosed skin blanches on application of pressure). It gives a slatey-grey hue.
4. Osteogenesis imperfecta—Only the sclera is blue. Due to thinness of the sclera, the choroid with its vessels give rise to blue tinge in the eyes. Cardinal features of this disease are fragile bones with multiple fractures, blue sclera, loose-jointedness and deafness.
5. Drug like amiodarone (used in ventricular arrhythmias) may produce bluish hue in the skin (ceruloderma).

Central plus peripheral (mixed) cyanosis :

1. CCF due to left-sided heart failure.
2. Acute myocardial infarction (cardiogenic shock) with acute LVF.
3. Rarely in polycythemia (red cyanosis).

What is differential cyanosis ?

1. Hands red (i.e., less blue) and feet blue—seen in PDA with reversal of shunt (differential cyanosis).

- Hands blue and feet red (i.e., less blue)—seen in coarctation of aorta with transposition of great vessels (reverse differential cyanosis).

* Intermittent cyanosis is seen in Ebstein's anomaly.

What is orthocyanosis ?

It is the development of cyanosis only in upright position due to hypoxia occurring in erect posture as a result of associated pulmonary arteriovenous malformations.

Table 25 : Bedside differentiation between central and peripheral cyanosis

Features	Central cyanosis	Peripheral cyanosis
1. Sites	1. As described above. Commonly seen in warm mucous membranes like tongue and oral cavity	1. As described above. Tongue remains unaffected
2. Handshake	2. Hand feels warm (due to increased blood flow)	2. Hand feels cold (due to diminished blood flow)
3. Application of warmth (by rubbing two palms) and cold (ice)	3. No change	3. Warmth—cyanosis decreased Cold—cyanosis increased
4. Application of pure oxygen for 10 minutes	4. Cyanosis may improve	4. No response
5. Clubbing and polycythemia	5. Usually present	5. Absent
6. Pulse volume	6. Normal or high (due to arteriovenous shunt) volume	6. Usually low volume
7. Dyspnoea	7. Patient is often breathless	7. No respiratory distress

* It is very difficult to diagnose cyanosis in the presence of polycythemia.

** Clubbing is absent in peripheral cyanosis and acutely developing central cyanosis.

Facts to be remembered :

- Cyanosis with clubbing—cyanotic congenital heart disease, pulmonary arteriovenous fistula, fibrosing alveolitis, extensive bronchiectasis, cystic fibrosis.
 - Cyanosis without clubbing—peripheral cyanosis and acutely developing central cyanosis.
- Clubbing without cyanosis—SBE, ulcerative colitis, normal healthy persons.

How to diagnose enterogenous cyanosis ?

- H/O ingestion of blue sweets or drugs (mentioned earlier).
- Skin and mucous membrane (tongue) are blue but the patient is not breathless.

You **are allowed to see only one site in cyanosis : what to do ?**

One should examine the tongue. **If the tongue is blue, the cyanosis must be central** but if the fingertips are blue, it may be either of central or peripheral in type.

Other sites looked for central cyanosis :

- Nasal mucous membrane examined by nasal speculum.
- Rectal mucous membrane seen by proctoscopy.
- Retina examined by ophthalmoscopy.

Both cyanosis and polycythemia are commonly present in :

- Cyanotic congenital heart diseases, and
- COPD.

How hypoxia and cyanosis are related ?

- Hypoxaemia without cyanosis—severe anaemia (Hb < 5 g/dl).
- Cyanosis with hypoxaemia—if reduced Hb is > 5 g/dl.
- Cyanosis without hypoxaemia—polycythemia vera (red cyanosis).

N.B. : After careful examination of the patient for cyanosis, one should examine for the presence of dyspnoea and clubbing. It should be remembered that a severely anaemic patient (Hb level below 5 g/dl) is possibly not cyanosed (though there is decreased O₂ saturation). For cyanosis to become clinically evident, there must be a minimum quantity of reduced Hb (at least 5 g/dl) in the blood perfusing the skin.

Case 38**JAUNDICE****Definition :**

It is defined as yellowish discolouration of the skin and mucous membrane due to excess amount of bilirubin present in the blood.

What is latent jaundice ?

The normal serum bilirubin level is 0.3-1.0 mg/dl. Clinical jaundice is evident when serum bilirubin crosses 3 mg/dl. Jaundice is latent, i.e., clinically non-evident (only detected by serum analysis) when the serum bilirubin level is in between 1-3 mg/dl.

Sites to be examined in a patient of jaundice :

Jaundice is always seen in bright natural daylight as it is not possible to detect light yellow colour in the presence of artificial light. **Take the patient in front of an open window** (never put a lighted torch) and look for :

1. ■ Upper bulbar conjunctiva (sclera is examined by retracting the upper eyelids upwards and asking the patient to look downwards - both eyes at a time).
2. Undersurface of tongue (sublingual mucosa).
3. Palate.
4. Palms and soles.
5. 'General skin surface.

Why the upper bulbar conjunctiva is selected ?

1. Sclera contains a lot of elastin (bilirubin has a strong affinity for elastic tissue), and
2. A white background is formed by sclera.

Differential diagnosis of jaundice :

1. Carotenaemia (carotenoderma)—Skin is yellow (mainly the palms and soles) but the sclera and mucous membrane are unaffected. The serum is also yellow in colour; the stool and urine are of normal colour.
2. Atabrine toxicity (previously used in malaria and tapeworm infestations)—Skin, urine and eyes (only the regions of the sclera exposed to light) are yellow.
3. Diffuse xanthomatosis—Skin takes a yellowish-orange tinge.
4. Muddy sclera—Exposed part of sclera looks dirty yellow; non-pathological.

Body secretions where bilirubin is found (in jaundice) :

1. Found in body fluids like CSF, joint fluid, cysts.
2. Absent from true secretions like tears, saliva, pancreatic juice.

* Scar tissue is rarely bilirubin-stained. Bilirubin has a strong affinity for elastic tissue (sclera, skin), nervous tissue (basal ganglia) and as it binds with proteins, it is more evident in exudates.

What is unilateral jaundice ?

It is seen :

1. In patients with hemiplegia.
2. In the presence of unilateral oedema.

Paralysed extremity and oedematous areas tend to remain less coloured as good circulation is necessary to stain the tissues.

3. Rarely, yellowish discolouration is seen in one eye when the other eye is 'artificial'.

* Right palm may look yellowish in health, who cooks at home (after using turmeric, specially in females).

Clinical classification of jaundice :

1. Hemolytic,
2. Hepato-cellular, and
3. Obstructive
 - a) Intrahepatic (medical).
 - b) Extrahepatic (surgical).

* Jaundice is described clinically as mild (< 6 mg/dl), moderate (6-15 mg/dl) and severe (>15 mg/dl).

** Jaundice may also be classified by chemical nature of bilirubin : conjugated and unconjugated hyperbilirubinaemia.

Features of haemolytic jaundice :

1. **Acholuric urine**, i.e., freshly passed urine is of normal colour as there is absence of bilirubin in urine (unconjugated urine is not water-soluble) but if the urine sample is kept for sometime, it turns dark yellow due to conversion of urobilinogen to urobilin (by oxidation).
2. Stool is high-coloured due to excess amount of stercobilinogen and stercobilin.
3. Jaundice is usually mild and there is **lemon-yellow tinge** of bulbar conjunctiva. Serum bilirubin is usually less than 6 mg/dl and predominantly of unconjugated variety.
4. Anaemia (mild, moderate or severe, according to the degree of haemolytic process).
5. Splenomegaly — Very characteristic (spleen is the slaughter house of RBC in haemolysis).
6. The patient may have typical facies (e.g., chipmunk facies in thalassaemia).
7. Reticulocytosis.

* In health, bilirubin can not be detected in urine. Urochrome and uroerythrin are the pigments which give normal urine its characteristic colour. In health, urinary urobilinogen (detected by Ehrlich's aldehyde test) and faecal stercobilinogen excretion per day are 1-3.5 mg and 40-280 mg respectively.

Features of obstructive jaundice :

1. Urine—Deep yellow or mustard oil-like colour (due to presence of conjugated bilirubin).
2. Stool—Pale or clay coloured (china-clay) with steatorrhoea (steatorrhoea means frothy, bulky, pale, offensive, soft, greasy stool which floats on the water of the pan, and is difficult to flush from the pan) due to absence of bile pigment in the stool.
3. Jaundice— Usually deep jaundice and the bulbar conjunctiva turns **greenish-yellow** due to oxidation of bilirubin to biliverdin (green). Jaundice is gradually progressive or fluctuating. Serum bilirubin is usually very high and predominantly of conjugated variety.
4. Generalised pruritus with scratch marks and shiny nails (bile acids irritate the free nerve endings).
5. Sinus bradycardia—Probably bile salts directly inhibit the sinoatrial node.
6. Xanthelasma around the eyes or xanthoma in the knees, buttocks may be seen due to hypercholesterolaemia.
7. Petechiae, purpura or ecchymosis are seen due to vitamin K deficiency (lack of bile salts produces vitamin K malabsorption as it is a fat soluble vitamin).
8. Gall bladder—May be palpable. This indicates the site of obstruction in the bile duct and is usually due to carcinoma of the head of pancreas (not due to choledocholithiasis)—according to **Courvoisier's law**.
9. Prolonged obstructive jaundice may produce osteomalacia, bone pain, bone fracture, or night blindness due to vitamin D, or A deficiency respectively (due to malabsorption or steatorrhoea). These type of bony changes in obstructive jaundice is known as 'hepatic osteodystrophy'.

10. Rarely, there may be hepatosplenomegaly. History of fever with chill and rigor, and pain abdomen may be present due to cholangitis. There may be abdominal pain in choledocholithiasis.

* Features of **chronic cholestasis** are pruritus with scratch marks, xanthoma, bruising and bone pain.

Features of hepato-cellular jaundice :

1. Urine—Yellowish.
2. Stool—High coloured and becomes pale if there is obstruction due to cellular (hepatocytes) oedema.
3. **Orange-yellow tinge** of the bulbar conjunctiva (conjugated > unconjugated bilirubin).
4. Anorexia, nausea, vomiting, fever with chill and rigor may be present before the appearance of jaundice (e.g. in viral hepatitis).
5. Tender hepatomegaly is frequent.
6. Variable pruritus.
7. There may be bleeding manifestations as a result of hepato-cellular failure.
8. There may be H/O affection of other members of the family or locality (viral hepatitis), drug intake (rifampicin) or poisoning (copper sulphate).

Bile salts and bile acids :

(A) BILE SALTS— Sodium taurocholate and sodium glycocholate.

(B) BILE ACIDS-

- a) Primary—Cholic acid and chenodeoxycholic acid.
- b) Secondary—Deoxycholic and lithocholic acid.
- c) Tertiary—Ursodeoxycholic acid.

What is Murphy's sign ?

Ask the patient of **acute cholecystitis** to breathe in deeply, and now try to palpate the gall bladder in sitting position. There is tenderness and catch in the breath at the height of inspiration with a mass felt there. This sign is not found in chronic cholecystitis. When the examination is performed with the patient lying, it is known as Moynihan's method.

What is Courvoisier's law?

Palpable, non-tender, smooth gall bladder in a patient of obstructive jaundice is due to neoplastic obstruction (usually due to carcinoma of the head of pancreas) of the common bile duct (CBD) and not due to impacted stone in the CBD. In case of choledocholithiasis, the gall bladder is usually small and contracted, and thus non-palpable due to repeated cholecystitis.

Table 26 : Differentiation between surgical and medical jaundice

Features	Extrahepatic obstruction (surgical)	Intrahepatic (medical) obstruction
(A) History :		
(1) Age	(i) Middle aged or old	(i) Usually young
(ii) Antecedent history	(ii) Previous attack, dyspepsia	(U) History of contacts, injections or blood transfusion, or nothing significant
(iii) Rate of development of jaundice	(iii) Slow	(iii) Rapid
(iv) Pain	(iv) Common (H/O colic)	(iv) Uncommon (usually ache over liver)
(v) Type of jaundice	(v) Fluctuates or persistent	(v) Acute onset, slow fall with recovery
(vi) Pruritus	(vi) Characteristically prominent feature	(vi) Often transient
(vii) Anorexia	(vii) Present but not so prominent	(vii) A prominent feature

Features	Extrahepatic (surgical) obstruction	Intrahepatic (medical) obstruction
(B) Examination:		
(i) Splenomegaly	(I) Rare	(i) May be present
(ii) Hepatomegaly	(H) Slightly tender hepatomegaly	(ii) Markedly tender hepatomegaly
(Ui) Palpable gall bladder	(iii) May be palpable due to carcinoma of the head of pancreas	(iii) Not palpable
(iv) Ascites	(iv) Usually absent	(iv) May be present in severe & prolonged obstruction
(v) Temperature	(v) May be raised (due to cholangitis)	(v) Raised initially
(vi) Anaemia	(vi) May be present	(vi) Usually absent
(C) Laboratory investigations :		
(I) WBC count in blood	(i) Increased or normal	(i) Diminished or normal
(ii) Differential count of WBC	(ii) Neutrophilia	(II) Increase in lymphocytes or eosinophils (in drug-induced jaundice)
(iii) Serum bilirubin	(iii) Usually 3-10 mg/dl; may be more	(iii) Varies with severity
(IV) AST and ALT	(iv) < 5 times of normal	(iv) > 5 times of normal
(v) Alkaline phosphatase	(v) > 3 times of normal	(v) < 3 times of normal
(vi) Ultrasonography or CT scan	(vi) May diagnose stones in CBD or carcinoma of the head of pancreas	(vi) Splenomegaly
(D) Therapeutic trial:		
'Prednisolone white wash' test	No response	Often +ve

Common examples of different types of jaundice in city hospitals :

(A) Haemolytic :

1. Thalassaemia.
2. Mismatched blood transfusion.
3. Snake bite (Viperidae group).
4. Malaria (specially falciparum malaria).
5. Rh incompatibility.
6. Primaquine or sulphonamide-induced (in G₆PD deficiency)

(B) Obstructive :

- a) Intrahepatic (medical) cholestasis :
 1. Cholestatic viral hepatitis.
 2. Chronic active hepatitis.
 3. Cirrhosis of liver (specially, primary biliary cirrhosis).
 4. Lymphoma or tuberculosis.
 5. Pregnancy in last trimester (recurrent cholestasis).
 6. Drugs—Chlorpromazine, chlorpropamide, oral contraceptives, erythromycin oestolate, methimazole, methyl testosterone, anabolic steroids.
 7. Secondary carcinoma of liver (jaundice is rarely seen in hepatoma).
 8. Dubin-Johnson syndrome.
- b) Extrahepatic (surgical) cholestasis:
 1. Gall stone impaction in CBD.
 2. Carcinoma of the head of pancreas, periampullary carcinoma.
 3. Carcinoma of the gall bladder.
 4. Enlarged glands at porta hepatis.

5. Stricture of CBD.

6. Sclerosing cholangitis (from inflammatory bowel disease), cholangiocarcinoma.

(C) Hepato-cellular :

1. Viral hepatitis (type A, B, C and E commonly).
2. Drugs — Rifampicin, INH, after halothane anaesthesia, paracetamol overdose.
3. Poisons — Copper sulphate.
4. Pregnancy — Acute fatty liver of pregnancy.
5. Alcoholic hepatitis, chronic hepatitis, cirrhosis of liver.
6. Weil's disease.
7. Right-sided heart failure.
8. Wilson's disease.

* A young person is more likely to have viral hepatitis (take H/O travel, alcohol abuse, male homosexuality, female prostitution, blood transfusion) whereas an elderly person with weight loss is more likely to suffer from carcinoma.

Diseases present as latent jaundice :

1. Mitral stenosis (i.e., passive venous congestion of liver).
2. Acute myocardial infarction.
3. Acute pulmonary thromboembolism.
4. Cirrhosis of liver.
5. Pernicious anaemia.
6. Acute pancreatitis.
7. Congestive cardiac failure.

Fluctuating jaundice :

1. Gilbert's syndrome.
2. Stone impaction in CBD (Charcot's biliary triad).
3. Periapillary carcinoma.
4. Haemolytic anaemias.
5. Chronic parenchymal liver disease (e.g. chronic active hepatitis).
6. Dubin-Johnson syndrome.

What is 'prednisolone white wash' test ?

If 30 mg of prednisolone (reduces cellular oedema) is given daily for 5 days to a patient of hepatocellular jaundice with features of obstruction (e.g., cholestatic viral hepatitis), there is a 40% fall in serum bilirubin. This test differentiates between medical and surgical obstructive jaundice (in a desperate situation) and gives positive result in medical jaundice. The test should not be used routinely. Before doing the test, hepatitis B and C markers must be negative.

Familial non-haemolytic hyperbilirubinaemias :

- | | |
|--|------------------------------------|
| 1. Gilbert's syndrome | I Unconjugated hyperbilirubinaemia |
| 2. Crigler-Najjar syndrome (type I and II) ' ' | |
| 3. Dubin-Johnson syndrome | I Conjugated hyperbilirubinaemia |
| 4. Rotor syndrome | J |

Differences between conjugated and unconjugated bilirubin :

	Unconjugated	Conjugated
1. Water solubility	0	+
2. Affinity for lipids	+	0
3. Renal excretion	0	+
4. van den Bergh reaction	Indirect	Direct

Fractionation of normal bilirubin :

Normal serum bilirubin level is 0.3-1.0 mg/dl.

(i) Conjugated fraction is 0.1-0.3 mg/dl, and (ii) Unconjugated fraction is 0.2-0.7 mg/dl.

Predominantly conjugated and unconjugated hyperbilirubinaemias :

(A) Conjugated hyperbilirubinaemia (with > 50% conjugated fraction) :

- (i) Viral or drug-induced hepatitis.
- (ii) Drug-induced cholestasis.
- (iii) Cholestatic jaundice of pregnancy.
- (iv) Cirrhosis of liver.
- (v) Extrahepatic biliary obstruction (e.g., stone, stricture etc.).
- (vi) Dubin-Johnson syndrome.
- (vii) Rotor syndrome.
- (viii) Secondary carcinoma of liver.

* Among steps of bilirubin metabolism (uptake, conjugation and excretion), excretion is the rate-limiting step and is usually affected most. Thus, conjugated hyperbilirubinaemia predominates in hepato-cellular diseases.

(B) Unconjugated hyperbilirubinaemia (usually with > 80% unconjugated fraction) :

- (i) Haemolysis.
- (ii) Ineffective erythropoiesis.
- (iii) Prolonged fasting (< 300 cal/day).
- (iv) Sepsis.
- (v) Gilbert's syndrome and rarely, Crigler-Najjar syndrome.
- (vi) Neonatal jaundice.

Hepatic disorders uncommonly associated with jaundice :

1. Hepato-cellular carcinoma (hepatoma).
2. Amoebic liver abscess.
3. Cystic disease of liver.

Can you detect jaundice at night ?

As previously discussed, jaundice should always be seen in broad daylight. But in a desperate situation, one has to examine the patient for jaundice at night remembering that in artificial light :

- Mild jaundice may be missed.
- Severely anaemic patient seems to have mild jaundice.
- Fluorescent or tube light gives yellow reflection in normal eye mimicking jaundice.

Medical causes of extrahepatic obstruction :

1. Sclerosing cholangitis in ulcerative colitis.
2. Obstruction by round worm in CBD.
3. Enlarged lymph nodes at porta hepatis in lymphoma.

* *Never put lighted torch inside the oral cavity to diagnose anaemia, cyanosis or jaundice.*

** Highest level of bilirubin is observed in hepato-cellular jaundice (may be in excess of 50 mg/dl). In cholestatic (obstructive) jaundice, bilirubin may reach upto 40 mg/dl and then remains around 30-40 mg/dl as a result of balance between renal excretion and conversion of bilirubin to other metabolites. In hepato-cellular jaundice, this balance is not seen rather high level of bilirubin may be explained by concomitant haemolysis and renal insufficiency.

Case 39

PULSE

Definition :

It is the expansion and elongation of the arterial wall imparted by the column of blood, and is passively produced by the pressure changes during ventricular systole and diastole. Commonly we examine the pulse in the radial artery in hand, which is one of the most accessible peripheral artery. Moreover, as soon as the hand (radial artery) of a patient is touched, it builds the doctor-patient relationship.

Pulse, blood pressure, respiration, temperature and level of consciousness are regarded as '**vital signs**' in clinical medicine.

Points to note in examination of pulse :

1. Rate.
2. Rhythm.
3. Volume.
4. Condition of the arterial wall.
5. Comparison between two radial pulses
6. Radio-femoral delay.
7. Any special character.
8. Palpation of other peripheral arteries.

N.B. : **Pulse should always be described under these eight points.** ‘Tension’ Is the indirect estimation of blood pressure. Estimation of tension by palpation is totally unreliable and thus, it is not included within the points under examination of pulse. Tension (i.e., blood pressure) should be accurately measured by sphygmomanometer.

How to examine the pulse ?

The radial pulse at the right wrist of the patient (present lateral to the flexor carpi radialis tendon) is generally examined with the pulp of three fingers (index, middle and ring finger). The patient’s forearm will be semipronated and the wrist slightly flexed.

The rate and rhythm are better palpated in the radial artery. To note the volume and character of the arterial pulse, one should examine the carotid artery (It is the nearest pulse to the aorta). Brachial pulse is felt to record blood pressure.

How to count the rate ?

Always count the beats for 15 seconds and multiply by four to get the pulse rate but it is worthwhile to remember that in a patient with arrhythmia, one must count the beats for at least one minute.

What are the variations in pulse rate ?

Definition of rate — Number of beats per minute. Normally it ranges between 60 to 100 beats per minute (average 72 beats per minute in an adult). Usually it remains 140 beats per minute at birth. SA node is the natural pacemaker of the heart (i.e., the normal cardiac rhythm is ‘**sinus rhythm**’).

(A) Tachycardia — Pulse rate is above 100 per minute. Normally it is found in children.

- a) Sinus tachycardia — Pulse rate is above 100 per minute where the impulse is originating from the SA node. The heart rate in sinus tachycardia varies between 100-160 per minute. The common causes are, (i) Exercise, emotion, excitement, intense pain, anxiety, and in children (ii) heart failure (iii) Thyrotoxicosis (iv) Severe anaemia (v) Pyrexia (vi) Shock (e.g, acute myocardial infarction) (vii) Myocarditis (viii) Acute haemorrhage (ix) Pregnancy (x) Hypoxia (xi) Drugs like salbutamol, nifedipine or atropine.
- b) Relative tachycardia — Read the section on Abnormal temperature’.
- c) Paroxysmal tachycardia — Pulse rate is above 160 beats per minute. It is divided into two types:
 - 1) Supraventricular (PSVT) [i.e., atrial (PAT) or nodal (PNT)].
 - 2) Ventricular (VT).

Common causes of paroxysmal tachycardia are rheumatic carditis, ischaemic heart disease (IHD), thyrotoxicosis, hypertensive heart disease, cardiomyopathy, WPW syndrome.

(B) Bradycardia—Pulse rate is below 60 per minute. It is commonly found in,

- | | |
|--|---|
| (i) Athletes, yoga, meditation or during deep sleep. | (vi) Hypothermia. |
| (ii) Myxoedema. | (vii) 2° heart block, complete heart block (CHB) |
| (iii) Obstructive jaundice. | (viii) Sick sinus syndrome (sino-atrial disease). |
| (iv) Increased intracranial tension. | (ix) Vasovagal attacks. |
| (v) Drugs like propranolol or digitalis. | (x) Severe hypoxia. |

* Athletes, yoga and meditation develop a high vagal tone but during sleep the sympathetic activity is reduced.

N.B. : Regular athletic training and myxoedema are the commonest causes of bradycardia.

- a) Sinus bradycardia—Pulse rate is below 60 per minute where the impulse is originating from the SA node. The common causes are : All the causes mentioned above in ‘bradycardia’ except complete heart block (in CHB, idioventricular rhythm occurs at the rate of 36 per minute).

N.B. : **Idioventricular rhythm**—when cardiac impulse arises from the ventricle at the rate of 36 per minute, the pulse rate at ventricular rhythm will be 36 (30-40) per minute, and is known as idioventricular rhythm; **nodal rhythm**—when the SA node fails, often the AV node originates impulse at the rate of 40-60 per minute resulting in a pulse rate of 40-60 per minute.

- b) Relative bradycardia — Read the section on ‘Abnormal temperature’.

* Tachycardia (Greek takhus = swift, kardia = heart)
Bradycardia (Greek bradus = slow, kardia = heart)

Bradycardia associated with convulsions :

1. Complete heart block (Stokes-Adams syndrome).

2. Increased intracranial tension due to meningoencephalitis, brain tumour, CVA.
3. Prior to development of coma in myxoedema.

Disproportionately rapid pulse in shock :

1. Acute myocardial infarction.
2. Septic shock.
3. Myocarditis.
4. Tachyarrhythmias.

What is pulse deficit (i.e., apex-pulse deficit) ?

Definition—it is the difference between the heart rate and the pulse rate. It is commonly found in atrial fibrillation and multiple ectopic beats. The method of detection of pulse deficit are :

1. First count the heart rate for one minute and then the pulse rate for next one minute (examination in two different cardiac cycles) — commonly practised method, or
2. One examiner counts the heart rate and the other examiner counts the pulse rate at the same time (using single cardiac cycle) for one minute (best method), or
3. One may put his stethoscope at the apex and simultaneously count the dropped beats in the pulse (using single cardiac cycle) for one minute.

* Pulse deficit is obtained in atrial fibrillation ($> 10/\text{min}$) and multiple ectopics ($< 10/\text{min}$).

What is an ectopic beat ?

In ectopic rhythm, the impulse arises from sites other than the SA node, and may arise from the atrial wall, AV node or ventricular wall. The ectopic beat is small, occurs prematurely and followed by a compensatory pause. The compensatory pause results in a 'missed' or 'dropped' beat in pulse.

Pulse felt at the wrist : Small pause followed by small beat, big pause followed by big beat.

Causes : Overindulgence of tea, coffee, cigarettes, alcohol; anxiety, dyspepsia; rheumatic, ischaemic, hypertensive, thyrotoxic and cardiomyopathic heart diseases; digitalis overdose.

Synonym of ectopic beat : Premature beat, extrasystoles.

* Ectopic beats occur as occasional or repeated irregularities superimposed on a regular pulse rhythm.

How can you modify ectopics (supraventricular) in a patient?

Ectopics can be abolished by vagotonic procedures, and vagolytic procedures increase the ectopics. When ectopic pace-maker is situated in the atrium (producing paroxysmal atrial tachycardia), vagotonic produces often terminate the attack. Vagotonic procedures have no effect on ventricular tachycardia.

Vagotonic procedures : Mechanical measures like *carotid sinus massage* (the patient lies flat with extended neck. Exclude occlusive carotid disease, i.e., there should not be any carotid bruit. Now put your left thumb first on the right side of the neck at the level of the upper border of thyroid cartilage and massage for 3-5 seconds at a time; next try on the left side with right thumb), self-induced gagging or vomiting, pressure over the eyeballs, Valsalva manoeuvre, coughing as well as breath holding, head lowering between the knees, stretching the arms and body; drug like digitalis.

Vagolytic procedures : Exercise; drugs like atropine and amyl nitrite.

Irregularities in the rhythm :

Rhythm is the spacing of successive beats (pulse wave) in time. Normal pulse is regular in rhythm and is known as **sinus rhythm**, because it is generated by the SA node. *Irregularities* are of two types :

1. **Regularly irregular** — i.e., irregularity comes at regular interval and is seen in extrasystoles, 2° heart block, sinus arrhythmia, pulsus bigeminus etc.
2. **Irregularly irregular** or completely irregular — i.e., irregularity between two pulse beats in every aspect (rate, rhythm, volume etc, i.e., totally chaotic). It is commonly seen in atrial fibrillation, multiple extrasystoles, atrial flutter with varying degrees of heart block etc.

What is sinus arrhythmia ?

Definition — It is the increase in pulse rate with inspiration and decrease in pulse rate with expiration. It is a physiological phenomenon commonly observed in children and athletes.

Mechanism — The increased amount of blood which comes in the left ventricle in expiration increases the stroke volume. This event immediately stimulates the baroreceptors (i.e., vagal stimulation) leading to slowing of the heart rate.

* Sinus arrhythmia may be absent in CCF and autonomic neuropathy.

Differentiation between multiple ectopics and atrial fibrillation (AF) :

Both the conditions present as irregularly irregular pulse. So, they have to be differentiated :

1. First count the pulse rate. If it is > 100 per minute, it is AF but if the pulse rate is < 100 per minute, it may be multiple ectopics or digitalised AF (treated case of AF).
2. The ectopic beat is a small one and occurs prematurely. So one may get the typical phenomenon of : small pause followed by small beat, big pause (compensatory pause) followed by big beat. If this typical cadence is felt, it is ectopic and not a case of atrial fibrillation (totally chaotic).
3. Now **count the pulse deficit**. If it is > 10, it is AF. If the pulse deficit is < 10, again it may be multiple ectopics or digitalised AF. To differentiate between the last two, following measures are adopted.
4. The patient is allowed to do mild exercise (if the condition permits). He will sit and touch the toes with his fingers, and will lie down again. This is done for 5-6 times successively. It is known that the heart of AF becomes more irregular after exercise. If after exercise the pulse deficit goes above 10, it is a case of digitalised AF and if the pulse deficit remains same (below 10) or diminished, the diagnosis of multiple ectopics becomes obvious.
5. Next the patient is examined for :
 - a) JVP—a-wave is absent in AF; a-wave is present in multiple ectopics.
 - b) S_j—Varying intensity of S_i in AF; no change in multiple ectopics.
 - c) Auscultation of the apex—If MS is the aetiology of AF, presystolic component of the murmur will disappear; no change of murmur in multiple ectopics.
6. ECG—'P' waves will be replaced by 'f' waves in AF; ectopics are easily diagnosed by ECG.
7. Fluoroscopy—Definite contraction of atria is absent in AF. Atria contracts normally in ectopics.

Irregular rhythm with normal heart rate :

1. Multiple extrasystoles.
2. Digitalised AF.
3. Sinus arrhythmia.

What is volume of the pulse ?

It is the amplitude of the pulse wave or the excursion felt at the wrist, and usually **reflects the width of pulse pressure** (systolic BP minus diastolic BP), which depends on two factors :

- (i) Stroke volume, and
- (ii) Compliance of the arteries.

The carotid, brachial or femoral arteries are more useful for assessing pulse volume and character than the radial pulse. Normal pulse pressure is 30-60 mm of Hg (i.e., normal pulse volume).

What are the changes in the pulse volume ?**(A) High volume pulse (i.e., pulse pressure > 60 mm of Hg) :**

1. Hyperkinetic circulatory states like.
 - (i) After exercise.
 - (ii) Severe anaemia.
 - (iii) Pyrexia.
 - (iv) Pregnancy.
 - (v) Aortic incompetence
 - (vi) Thyrotoxicosis.
 - (vii) Arteriovenous communications like patent ductus arteriosus (PDA), Paget's disease etc.
 - (viii) Chronic cor pulmonale.
 - (ix) Hepato-cellular failure.
 - (x) Beri-beri.
2. Atherosclerosis (the arteries are rigid, i.e., less compliant; so there is wide pulse pressure as the systolic BP is high).
3. Complete heart block or bradycardia due to any cause.

* In aortic incompetence : systolic BP ↑ and diastolic BP ↓; atherosclerosis : systolic BP ↑ and diastolic BP normal; hyperkinetic circulation : systolic BP ↑ and diastolic BP near normal.

(B) Low volume pulse (pulsus parvus; pulse pressure < 30 mm of Hg) :

1. Shock due to any cause e.g., acute myocardial infarction, massive haemorrhage, hypovolaemia or septicemic shock (as a result of poor vascular tone).
2. Severe aortic stenosis.
3. Tight mitral stenosis.
4. Pericardial effusion.
5. Constrictive pericarditis.
6. Congestive cardiac failure.

What is a thready pulse ?

This is a low volume pulse with rapid pulse rate. This type of pulse is seen in peripheral circulatory failure, e.g., cardiogenic shock, haemorrhage or dehydration.

* 'jerky pulse' is felt in idiopathic hypertrophic subaortic stenosis (IHSS).

How to assess the condition of the arterial wall ?

Normally the arterial wall is impalpable and *may be palpable in old age* due to arteriosclerosis. The artery becomes tortuous, thickened and feels like a cord in arteriosclerosis which is known as "Monckeberg's medial sclerosis" (calcification of the medial coat of large arteries). This is an age-related change; remember, atherosclerosis occurs in intima and often associated with occlusive arterial disease. In the presence of thickened and tortuous artery, one may observe locomotor brachialis. Condition of the arterial wall is assessed by :

- a) Compress the brachial artery above the elbow by ball of the left thumb (i.e., making a bloodless column) and now roll the radial artery over radius by index and middle fingers of the right hand, Or
- b) First place the index and middle fingers of both the left and right hand over the radial artery side by side and exsanguinate the artery by moving the two middle fingers in opposite direction. The radial artery is now rolled over the radius by two index fingers.

Causes of absent radial pulse :

1. Anatomical abnormality.
2. Severe atherosclerosis.
3. Takayasu's disease.
4. Embolism in the radial artery'.
5. [Death].

* Raynaud's disease doesn't affect the peripheral pulse because it is a disease of arterioles.

Inequality between two radial pulses (radio-radial delay) :

Simultaneously palpate both the radial arteries by both of your hands, using your left hand for patient's right hand and vice-versa for the other hand. Causes of inequality or radio-radial delay are :

1. Normal anatomical variations.
2. Thoracic inlet syndrome e.g., cervical rib.
3. Aneurysm of the arch of aorta.
4. Pre-subclavian coarctation.
5. Supravalvular aortic stenosis (congenital).
6. Pulseless disease (Takayasu's disease).
7. Peripheral embolism or atheromatous plaque.
8. Atherosclerosis of aorta.
9. Pressure over axillary artery by lymph nodes.
10. Iatrogenic—Blalock-Taussig shunt operation in Tetralogy of Fallot.

Causes of radio-femoral delay :

For detection of radio-femoral delay, standing on the right side of the patient, one should palpate the radial artery with the left hand and femoral artery with the right hand simultaneously. **Normally there is no radio-femoral delay** (in health, radial and femoral pulsations are felt equally and synchronously) but it should be remembered that the pulsation of arteria dorsalis pedis comes 0.02 to 0.03 seconds later than the radial artery. In radio-femoral delay, the femoral pulse is of small volume and occurs after the radial pulse. Causes of radio-femoral delay are:

1. Coarctation of aorta (important bedside diagnostic clue in a young hypertensive).
2. Atherosclerosis of aorta.

3. Thrombosis or embolism of aorta.
4. Aortoarteritis.

Basic bedside features of coarctation of aorta :

1. A male patient with headache, claudication, palpitation, anginal pain or cold extremities.
2. The upper extremity and thorax may be more developed in comparison to lower extremities.
3. Palpation of pulses may reveal **radio-femoral delay** (symmetrical reduction and delay of femoral pulses in comparison to radial pulses). All the pulses like radial, carotid, brachial, femoral, popliteal and *arteria dorsalis pedis* should be examined in details.
4. Prominent suprasternal and carotid pulsation.
5. Collateral pulsation are present (seen as well as felt) in axilla, trunk and infrascapular area (Suzman's sign). Suzman's sign (dilated, tortuous and pulsatile arteries) is best elicited when the patient stands and bends forward with arms hanging down at sides.
6. **Systemic hypertension** (upper extremity high BP with low or normal BP in lower extremity).
7. Bruit over the collaterals. 'Cork-screw' appearance of retinal arteries (fundoscopy).
8. Left ventricular type of cardiac enlargement (heaving apical impulse); a systolic murmur may be heard over the anterior chest and back. Continuous murmur may be present over collaterals.
9. Clinical associations : Bicuspid aortic valve, PDA. VSD, berry aneurysm, polycystic kidneys, Turner's syndrome.
10. [Rib notching from 3rd to 8th rib in chest X-ray — known as Dock's sign].
11. [Barium swallow of oesophagus shows double indentations due to pre- and post-coarctation dilatation—'3 sign' or 'reverse E sign'.]

* Post-subclavian type is the commonest variety of coarctation of aorta.

Examination of other peripheral pulses :

1. Ulnar artery : At the wrist on medial side where it crosses the distal end of radius.
2. Brachial artery : Palpated by the thumb (left thumb for left arm and vice-versa for right arm) at or just above the elbow (just medial to the tendon of biceps muscle).
3. Subclavian artery : Ask the patient to shrug his shoulders. The artery is felt from behind by pressing the index finger downwards, from above the middle of the clavicle.
4. Carotid artery : Palpated with the thumb (left thumb for right carotid and vice-versa), medial to sternomastoid and at or just below the level of thyroid cartilage. Pressure over the carotid sinus may sometime induce syncope and this is why carotid artery should always be palpated one by one with the patient lying in bed. Dancing carotids in AI, slow upstroke in AS and jerky carotids are characteristic of HOCM.
5. Femoral artery : Lies midway between anterior superior iliac spine and pubic tubercle, at the level of the groin or inguinal ligament.
6. Popliteal artery : The patient lies supine with semiflexed knee. The fingertips of both hands are pressed in the middle of the popliteal fossa while both thumbs rest on tibial tuberosity. It may be palpated in prone position with knee partly flexed.
7. Posterior tibial artery : Lies approximately in the midway between the medial malleolus and heel; better palpated after inversion of the foot as it causes relaxation of the flexor retinaculum.
8. Arteria dorsalis pedis : Palpated lateral to the extensor hallucis longus tendon on the proximal part of dorsum of the foot; it may be absent in about 10% people. Before palpation, make the extensor hallucis longus tendon prominent by asking the patient to extend his great toe against resistance.

Describe the normal arterial pulse (adult) :

The rate is 72 per minute. The beats are regular in rhythm, and normal as well as equal in volume. There is no inequality between the two upper limbs, and the upper and the lower limbs. The condition of the arterial wall is normal (neither thickened nor tortuous) and the pulses are of normal **character (catacrotic pulse)**. All the peripheral pulses are symmetrically palpable.

* Character of pulse is described in the section on 'Water-hammer pulse'.

** Throbbing carotids in aortic incompetence is known as 'Corrigan's pulse'.

*** Examination of the pulse is one of the most ancient and time-honoured practice of the medical profession. Try to comment on pulse deficit while describing the pulse of atrial fibrillation.

**** Pulse becomes faster (rapid) after a meal, after smoking, during menstruation and in the evening.

Case 40

WATER-HAMMER PULSE

Define 'character' of pulse :

The volume, waveform and some special features (e.g., collapsing nature) altogether give rise to character of the pulse, which is often helpful to clinical diagnosis of specific diseases or disorders.

Different 'characters' of pulse :

- | | |
|------------------------|--|
| 1. Anacrotic pulse. | 5. Pulsus alternans. |
| 2. Dicrotic pulse. | 6. Pulsus bigeminus. |
| 3. Water-hammer pulse. | 7. Pulsus paradoxus. |
| 4. Pulsus bisferiens. | 8. Thready pulse, jerky pulse, bounding pulse. |

What is catacrotic pulse ?

The normal arterial pulse is known as catacrotic pulse which is described in the section on 'Pulse'.

It has following waves :

- Percussion wave (P, the rapid upstroke in pulse, and is produced by ejected blood in arterial system).
- Tidal wave (T, and is generated along the arterial wall).
- Dicrotic notch and dicrotic wave (in downstroke of pulse; due to elastic recoil of the vessel).

The wavy pattern is not felt in health since it is obliterated by normal vascular tone.

What is pulse pressure and mean pressure ?

Pulse pressure is the difference between systolic and diastolic BP (roughly reflects the volume of the pulse). Mean pressure is approximately the arithmetic mean of diastolic and systolic pressure, and is calculated by diastolic pressure plus 1/3rd of pulse pressure.

What is a bounding pulse ?

It is commonly seen in hyperkinetic 'circulatory states where the pulse volume is high (due to high pulse pressure) with increased blood flow.

What is water-hammer pulse (or high volume collapsing pulse) ?

This is characterised by :

1. High volume pulse,
2. Sharp rise,
3. Ill-sustained, and
4. Sharp fall.

A high volume pulse gives the water-hammer character when the pulse pressure is at least above 60 mm of Hg. A **collapsing pulse** usually occurs where there is rapid run-off of blood from the aorta or the arterial system. The collapsing nature is often reliably detected by palpation of the carotid artery.

* All collapsing pulses are of high volume but all high volume pulses are not necessarily collapsing.

Demonstration of water-hammer pulse :

Palpate the wrist in such a way that your webs fall on the radial artery and rest of the palm lies over the ulnar artery. Examine the volume of the pulse (summation of both radial and ulnar artery) for few seconds. Now elevate the whole upper limb suddenly above the level of the heart (may give a support in the elbow to prevent its flexion) and try to recognise any changes in the volume of the pulse. In water-hammer pulse, the volume increases from the basal level (i.e., volume at the beginning of the examination) after elevation of the upper limb, and the pulse strikes the palpating palm with a rapid and forceful jerk. Abrupt downstroke of the pulse produces the collapsing feel.

For examination of the pulse in this way, one should stand within the 'angle' formed between the patient's body and the said upper extremity. The right-sided pulse should be examined by the right hand while standing on the right side, and vice-versa for the left. Observe the increase in pulse volume, sharp rise and the sharp fall.

Why the name 'water-hammer' ?

It is termed after a 19th century Victorian toy called 'water-hammer'. This is a peculiar toy where a sealed glass cylinder is half filled with water and half with vacuum (two ends being closed). If the toy is suddenly placed upside down, the column of water strikes the other end of the cylinder with a blowing

sound. This is why the term water-hammer has been coined in a high volume collapsing pulse where the pulse strikes the fingers like the thud of a hammer.

Causes of water-hammer pulse :

The causes are mentioned in 'high volume pulse' in the section on 'Pulse'. One should remember **aortic incompetence (AI)** as the first cause of water-hammer pulse and then the others.

Why this type of pulse is seen in AI ?

(A) In AI, the left ventricular stroke volume is high (so the systolic pressure is high) and this is responsible for 'sharp rise' or rapid upstroke in the pulse.

(B) The collapsing character is due to,

1. Diastolic leak back into the left ventricle from aorta.
2. Rapid run-off to the periphery as a result of *low systemic vascular resistance* (the increased cardiac output stimulates the baroreceptors in the aortic arch and the result is reflex vasodilatation of the peripheral vessels into which the blood flows rapidly).

N.B. : 1 and 2 of (B) are responsible for low diastolic pressure and thus in AI, there is a high pulse pressure. These factors are responsible for 'sharp fall' too.

Why do you elevate the arm ?

Mechanisms of action :

1. Aided by the gravity, there is fall of blood column (resulting in vasodilatation) and thus, it helps to reduce the diastolic pressure more. So the pulse pressure widens, or
2. It may be so that the artery palpated becomes more in the line of aorta after elevation of the arm, and thus allows direct systolic ejection and diastolic backward flow.

Other parts to be examined in a patient with water-hammer pulse :

1. Count the rate (bradycardia in complete heart block; tachycardia in thyrotoxicosis).
2. Examine the condition of the arterial wall (for atherosclerosis).
3. See the facies for exophthalmos or examine for tremor (thyrotoxicosis).
4. Record the surface temperature (pyrexia).
5. Examine for anaemia (severe anaemia).
6. Examine for jaundice (cirrhosis with hepato-cellular failure).
7. Examine the chest for emphysema (chronic cor pulmonale).
8. One should not forget to auscultate the aortic and neo-aortic area for an early diastolic murmur (AI).

* Lastly, one should examine the patient for capillary pulsation, digital pulsation, carotid dance, pistol shot sound which are usually associated features of water-hammer pulse.

Anacrotic pulse :

It is a low volume pulse with an upstroke felt in the ascending limb. Anacrotic pulse is found in severe valvular aortic stenosis (AS). Typically the pulse in aortic stenosis is known as pulsus '**parvus et tardus**' (slow rising low volume pulse)— '**parvus**' means low volume and '**tardus**' means slow or late. One may feel carotid shudder in the presence of anacrotic pulse. Pulse of AS is also known as 'plateau pulse'.

Dicrotic pulse :

When an upstroke is felt in the descending limb of the pulse wave, it is known as dicrotic pulse. The dicrotic wave is felt due to hypotonia of the vessel wall. This is also a *low volume* pulse due to very low stroke volume with decreased peripheral resistance, and is found in :

- a) Second week of typhoid fever (possibly related to circulating vasculotoxins).
- b) Endotoxic shock.
- c) Hypovolaemic shock.
- d) Diffuse myocardial disease.

What is pulsus bisferiens ?

1. It is a high volume double-beating pulse (single pulse wave with two peaks in systole).
2. Method of palpation : best palpated in big arteries like brachial and carotid arteries. Try to press and occlude the brachial artery. Now, slowly release the pressure and feel the double-beating nature of the pulse.
3. The first lift is due to 'percussion wave' (P) and the second lift is due to 'tidal wave' (T). It is said that if $P > T$, then $AI > AS$; and if $T > P$, usually $AS > AI$. Thus, we can determine the dominant lesion.

4. It is commonly found in,
 - a) Combined AS and AI (commonest),
 - b) Isolated AI, and
 - c) Hypertrophic obstructive cardiomyopathy (HOCM).
 5. Pathophysiology—
 - a) Due to 'ventury effect' within the left ventricle.
 - b) Another view explains that the 'dip' in the pulse is felt due to energy dissipation in the production of a loud systolic murmur of AS (Bernoulli effect).
 - c) The first component is due to large volume of blood ejected in systole and the second component is due to elastic recoil in the arteries (best reasoning).
- * Three different causes of **double-beating pulse** are (i) bisferiens, (ii) anacrotic, and (iii) dicrotic pulse.

Pulsus alternans :

This is rare. When the alternate pulse waves are weak i.e., of low volume (*the rhythm remains regular in contrast to ectopics*) in a patient with acute left ventricular failure, pulsus alternans is said to be present. **It is better demonstrated in radial arteries.** In a patient with LVF (severe myocardial failure), some ventricular muscle fibres are healthy and some are degenerated and thus, produces normal and weak beat respectively (defective electromechanical coupling). One should search for gallop rhythm and basal crepitations when pulsus alternans is felt. It is due to prolonged recovery time of damaged myocardium and indicates a poor prognosis.

The compensatory pause of ectopics are absent here.

Pulsus bigeminus :

Clinically, two beats and a pause thereafter recur repeatedly in a regular fashion (bigeminy means twins). The second beat is an ectopic and thus, there is a pause after it. It is commonly found in digitalis toxicity and 3 : 2 heart block. In pulsus trigeminus, three beats and a pause recur in a regular fashion.

Pulsus paradoxus :

Here, the pulse volume decreases with inspiration and increases with expiration (not truly the opposite of sinus arrhythmia which denotes the changes in pulse rate only). The '**paradox**' is that the heart sounds may still be audible on auscultation over the apex at a time when no pulse (pulse wave may totally disappear in inspiration) is palpable at the radial artery. It is commonly seen in,

1. Acute severe asthma.
2. Cardiac tamponade (rapidly developing pericardial effusion).
3. Chronic constrictive pericarditis.
4. Chronic obstructive pulmonary disease (COPD).
5. Sometimes, SVC syndrome.
6. Restrictive cardiomyopathy.

The probable mechanisms are :

1. Intrapericardial pressure rises more during inspiration (as a result of traction on the pericardium) which in turn diminishes cardiac output by causing obstruction in venous return (in cardiac tamponade). Increased intrapericardial pressure in cardiac tamponade also impedes diastolic filling.
2. Anti-Bernheim effect—During inspiration, more blood comes in the right ventricle which pushes the interventricular septum to the left side and thereby diminishing the left ventricular cavity, and resulting in low cardiac output. During acute severe asthma and other causes mentioned above, the degree of negative pressure generated during inspiration is exaggerated. So, more blood is sucked in the right heart, resulting in pulsus paradoxus.

* Pulsus paradoxus is a misnomer and is actually an exaggeration of normal physiological phenomenon. In health, there is a fall in systolic BP in inspiration which is < 10 mm of Hg (so, not detectable normally) but in pulsus paradoxus, there is an exaggerated inspiratory fall in systolic BP > 10 mm of Hg.

Pulses confirmed by sphygmomanometer :

1. Pulsus paradoxus—Systolic BP is more in expiration than in inspiration by > 10 mm of Hg.
2. Water-hammer pulse—Pulse pressure is usually greater than at least 60 mm of Hg.
3. Pulsus alternans—When the strong beats are heard during measurement of systolic BP (initial part of measurement), the pulse rate remains half of the actual rate (as weak beats do not reach

the radial artery). With gradual lowering of the mercury column, the weak beats are also heard and thus, the pulse rate doubles i.e., returns to the actual pulse rate (Gallavardin sign).

Other uses of sphygmomanometer :

1. To measure the blood pressure (principal use of the instrument).
2. Hess' capillary fragility test—See the section on 'Haemorrhagic spots'.
3. Latent tetany—When the pressure is raised above the systolic BP for 3 minutes, typical carpal spasm appears (main d' accoucheur) and is known as Trousseau's sign.
4. To assess the respiratory reserve—Blow the mercury column (by placing the mouth to the inlet tube) upto 40-50 mm of Hg and try to hold it at this level.
5. Diagnosis from recording of BP of lower limb : Lower limb systolic BP > upper limb systolic BP and if the difference is above 20 mm of Hg, it is known as Hill's sign, which is diagnostic of AI. Lower limb systolic BP < upper limb systolic BP happens to occur in coarctation of aorta.
6. To draw venous blood.
7. Previously used as a rotating tourniquet in a patient of LVF (for reduction of venous return) Keep the cuff inflated in all the limbs at 80 mm of Hg (i.e., at diastolic pressure) for 15 minutes.
8. To draw blood during blood donation.

Describe the pulse in complete heart block (CHB) :

1. Rate—36 to 40 per minute (bradycardia).
2. Rhythm—Regular.
3. Volume—High.
4. Condition of the arterial wall—May be thickened (as the patients are of advanced age).
5. Neither radio-radial nor any radio-femoral delay.
6. Character—May have water-hammer character.
7. All the peripheral pulses are palpable.
8. Special character—It is a 'fixed' pulse i.e., no alteration in pulse rate after exercise, pyrexia or injection of atropine.

In CHB carotid artery shows carotid dance at the rate of 36-40 per minute (ventricular rate), and internal jugular veins reflect atrial pulsation at the rate of 72 per minute with cannon waves appearing from time to time. There is beat to beat variation in the BP. BP is of high systolic and normal diastolic pressure. There is varying intensity of S₁ (cannon sound); rarely a mid-diastolic murmur may be audible (Rytand's murmur).

Case 41

ABNORMAL TEMPERATURE

Normal body temperature and its variations :

Fever is only a symptom of disease which alerts the physician to the underlying abnormality.

1. **Normal**- 98°F-99°F (with a diurnal variation of 1-5°F; the temperature is lowest in the morning and is highest in between 4-6 PM).
2. **Subnormal**- below 98°F.
3. **Pyrexia or febrile**- above 99°F.
4. **Hypothermia**- below 95°F.
5. **Hyperpyrexia**-above 106.7°F.

* Though varies from person to person, recent studies reveal that oral AM temperature of > 98.9°F or a oral PM temperature of > 99.9°F would define a fever or pyrexia.

Methods of taking body temperature :

1. **Oral**- The thermometer is placed under the tongue and the patient breathes through the nose with lips firmly closed. It reflects the body-core temperature.
2. **Axillary** or groin- Placed in the armpit or folded groin; generally 0.5-1 °F lower than oral temperature. Axilla, groin and natal cleft should preferably be avoided in adults.
3. **Rectal**- Most reliable as well as accurate, and it is 0.5-1 °F higher than the oral temperature.

N.B. : Thermometer is kept at least for 2 minutes in all the sites and the rectal > oral > axillary temperature

* Recently, a tympanic membrane thermometer (i.e.. electronic thermometer used in the ear) is used for fast and accurate reading of core temperature.

Which site will you prefer in infants ?

The axilla, or groin with thigh flexed over the abdomen (also convenient in an unconscious patient).

If not mentioned, which site is preferred for recording of temperature ?

Oral; if the oral temperature crosses 99°F, the patient is febrile. In axilla, when the temperature is above 98-6°F or 37°C (the arrow is marked here in the clinical thermometer), it is said that the patient is running temperature. Oral temperature does not change with sweating or vasoconstriction (axillary temperature may change). Majority of common people are well conversant with Fahrenheit scale.

Contraindications of taking oral temperature :

1. Infants and children.
2. Mentally dull persons, e.g., Down's syndrome.
3. During convulsions or H/O recurrent fits.
4. After taking hot tea, hot coffee or chewing tobacco; smoking.
5. Inflammation within the oral cavity, e.g., stomatitis, ulcer in the mouth, impacted wisdom teeth.
6. During severe dyspnoea.
7. Unconscious or restless patients.
8. Trismus (tetanus, tetany).
9. Paralysis of the tongue; Bell's palsy.
10. In mouth breathers.

Causes of hypothermia :

1. Myxoedema coma.
2. Prolonged exposure to cold (in street beggars, accidental in elderly, or in mountaineering).
3. Peripheral circulatory failure due to any cause.
4. Enteric fever if associated with haemorrhage or perforation : commonly occurs in the 3rd week.
5. Hypoglycaemia.
6. Panhypopituitarism; adrenal insufficiency.
7. Artificial hypothermia induced in open heart surgery.
8. Near-drowning.
9. Alcohol intoxication; sedatives and hypnotics overdose.
10. Autonomic dysfunction.

* Always confirm hypothermia with rectal temperature.

Causes of hyperpyrexia :

- | | |
|---------------------------|-------------------------------------|
| 1. Malaria. | g Thyroid storm. |
| 2. Septicaemia. | 9. Neuroleptic malignant syndrome |
| 3. Encephalitis. | (haloperidol-induced). |
| 4. Pontine haemorrhage. | 10. Dhatura poisoning. |
| 5. Lobar pneumonia. | 11. Rabies. |
| 6. Heat stroke. | 12. Serotonin syndrome (SSRI or MAO |
| 7. Malignant hyperthermia | inhibitors-induced). |
| (halothane-induced). | |

Causes of 'aseptic fever' :

- | | |
|---|---------------------------------|
| 1. Heat stroke. | 6. Thyroid storm. |
| 2. Lymphoma, leukaemias or | 7. Acute myocardial infarction, |
| disseminated malignancy. | 8. Gout. |
| 3. Collagen vascular disease e.g., SLE. | 9. Porphyria. |
| 4. Pontine haemorrhage. | 10. Crush injury. |
| 5. Drug fever e.g., rifampicin or | 11. Radiation sickness, |
| sulphonamides. | 12. Over-atropinisation. |

Fever with disturbed consciousness :

1. Encephalitis.
2. Cerebral malaria.
3. Meningitis.
4. Pyrexia complicated by metabolic disorders e.g., hypoglycaemia.
5. Typhoid state.
6. Brain abscess.
7. CVA with fever (e.g., chest infection)
8. Rarely in dhatura poisoning.

What is 'hectic' temperature ?

There is a big swing in the temperature which rises with chill and rigor, persists for few hours and suddenly falls with profuse sweating. The common causes are :

1. Pent-up pus anywhere in the body (lung abscess, liver abscess, empyema thoracis, empyema of gall bladder, subdiaphragmatic abscess).
2. Septicaemia or pyaemia.
3. Rarely in advanced tuberculosis.

Causes of fever with chill and rigor :

1. Malaria.
2. Urinary tract infection.
3. Pent-up pus anywhere in the body (e.g., lung abscess).
4. Septicaemia or pyaemia.
5. Cholangitis.
6. Subacute bacterial endocarditis.
7. Thrombophlebitis.
8. Acute pyelitis or acute pyelonephritis.
9. Acute lobar pneumonia (e.g., pneumococcal).
10. Agranulocytosis.
11. Pyrogen reaction after fluid infusion or blood transfusion.
12. Filariasis.

Pathogenesis of chill and rigor :

It should be looked upon in the setting of 'thermostat'. When the temperature is rising to a higher level, heat is being conserved, the cutaneous vessels are constricted (so the patient feels cold) and the patient may even shiver violently. The shivering is known as 'rigor'.

When the higher temperature is reached, heat loss starts and the cutaneous vessels dilate for dissipation of heat. The patient feels hot and sweating starts.

Normal temperature is maintained in health by regulating a balance between heat gain and heat loss, governed by the hypothalamus.

What is meant by 'fall by crisis or lysis' ?

Fall of temperature is also known as defervescence. They are :

- I. **Crisis**—When the raised temperature falls to normal or subnormal level very quickly within 6-12 hours and is associated with severe sweating, it is known as fall by crisis. This is usually seen in,
 1. Acute lobar pneumonia.
 2. Enteric fever complicated by intestinal haemorrhage or perforation.
 3. Adrenal crisis (seen in meningococcal meningitis).
 4. Septicaemic shock.
 5. Dengue.
- II. **Lysis**—When the temperature falls gradually in steps over several days, it is known as fall by lysis. It is usually seen in,
 1. Uncomplicated enteric fever.
 2. Rheumatic fever.
 3. Acute bronchopneumonia.

N.B. : Most of the raised temperature falls by lysis. Use of antipyretics may precipitate fall by crisis.

What do you mean by 'periodic fever' ?

It is the periodic attacks of pyrexia alternating with a period of apyrexia, and is commonly seen in :

1. Hodgkin's disease (Pel-Ebstein fever).
2. Brucellosis.
3. Relapsing fever.
4. Malaria.
5. Rat bite fever.
6. Dengue.

Diagnostic clues in fever :

- Toxic look—Typhoid, septicaemia, miliary tuberculosis, typhus.
- Marked perspiration—Septicaemia, rheumatic fever, amoebic liver abscess, brucellosis.
- Severe myalgia—Influenza, dengue, chikungunya, leptospirosis.
- Purpura—Acute leukaemias, septicaemia.
- Convulsions—Febrile (in children), meningitis, cerebral abscess, tuberculoma.
- Lymphadenopathy Tuberculosis, lymphoma, leukaemia, secondary syphilis, SLE, AIDS.

What is pyrexia of unknown origin (PUO) ?

It is also known as fever of unknown origin (FUO).

Definition (by Petersdorf and Beeson. 1961) :

1. Fever higher than 101°F on several occasions.
2. A duration of fever for more than 3 weeks.
3. Failure to reach a provisional diagnosis after one week of inpatient investigations.

Diseases which may initially present as PUO :

1. Lymphomas, leukaemias, multiple myeloma and carcinoma (specially of lung, liver and kidney).
2. Collagen vascular diseases, e.g., SLE, rheumatoid arthritis. Still's disease, polyarteritis nodosa.
3. Tuberculosis, SBE, subphrenic abscess, liver abscess, brucellosis, fungal infection. AIDS.
4. Factitious fever, drug fever, habitual hyperthermia, use of immunosuppressive drugs.

Recently PUO has been classified into 4 different types (Durack and Street, 1991) •

1. Classic PUO (as previous one except the time frame which is 3 outpatient visits or 3 days in hospital without determining the cause, or one week of intelligent and invasive ambulatory investigations)—e.g., hidden infection, obscure malignancy, collagen vascular diseases.
2. Nosocomial PUO (hospital-acquired)—e.g., septic thrombophlebitis.
3. Neutropenic PUO (when neutrophil count is < 500/mmr³)—e.g., perianal infection, candidiasis.
4. HIV-associated PUO (e.g., tuberculosis, NHL, drug fever).

N.B. : Diagnosis of PUO requires 'repeated' physical examination, intelligent interviewing the patient and specific investigations.

* Fever persisting for > 2 weeks in loosely termed as 'prolonged fever'.

When the rectal temperature is preferred ?

1. Possible hypothermia.
2. In collapsed, comatosed or elderly patients.
3. Invalid patients.
4. Cholera patients (surface temperature is subnormal but rectal temperature is raised), and
5. In algid malaria.

N.B. : The rectal temperature is recorded with a special 'low-reading' thermometer. It is stouter than the clinical thermometer, with a rounded bulb (to reduce the risk of injury) and is graduated upto 90°F.

Relative bradycardia and relative tachycardia :

It is known that with per degree (°F) rise of temperature in an adult, the pulse rate is increased by 10 beats/minute. In children, the rise may be upto 12-15 beats/minute.

(A) **Relative bradycardia** In this condition, the increase in pulse rate is less than 10 beats/minute with per degree (°F) rise of temperature. For example, the pulse rate in relative bradycardia will be about 80/minute, when the temperature is increased by 2°F (actually it should be 92/minute) It is seen in :

- (i) Any viral fever (e.g., in yellow fever - known as *Faget's sign*; dengue).
- (ii) First week of enteric fever.
- (iii) Sometimes in pyogenic meningitis.
- (iv) Brucellosis, psittacosis, Weil's disease.

(B) **Relative tachycardia**— Here, the increase in pulse rate is more than 10 beats/minute with per degree (°F) rise of temperature. For example, the pulse rate will be about 120/minute, when the temperature is increased by 3°F (actually it should be 102/minute). It is seen in :

- (i) Acute rheumatic carditis.

- (ii) Tuberculosis.
- (iii) Diphtheritic myocarditis.
- (iv) Polyarteritis nodosa.

Types of fever :

There are three classical types of fever :

- I. **Intermittent-** Fever is present only for several hours and **always touches the baseline** sometime during the day. It has three subdivisions :
 - (i) Quotidian-The paroxysm of fever occurs daily (i.e., daily rise and daily fall). Examples are,
 - a) Double infection of *P. vivax*.
 - b) Pent-up pus anywhere in the body.
 - c) Tuberculosis.
 - d) Urinary tract infection.
 - e) Septicaemia.
 - (ii) Tertian- The paroxysm occurs on alternate days, which is found in,
 - a) Benign tertian malaria (commonly by *P. vivax* and rarely by *P. ovale*).
 - b) Malignant tertian malaria (*P. falciparum*).
 - (iii) Quartan- When two days intervene between consecutive paroxysmal attacks, it is quartan pyrexia and is found in quartan malaria (*P. malariae*) which is rare in India.
- II. **Continued-** Fever does not fluctuate more than 1°C (1 -5°F) during the twenty four hours period and **never touches the baseline**. The examples are,
 - a) Lobar pneumonia.
 - b) Second week of enteric fever.
 - c) Miliary tuberculosis.
 - d) Meningococcal meningitis.
 - e) Rheumatic fever.
- III. **Remittent-** Daily fluctuation of fever is more than 2°C (3°F) during the twenty four hours period and it **never touches the baseline**. The common examples are,
 - a) Amoebic liver abscess.
 - b) Sometimes in urinary tract infection.
 - c) Third week of enteric fever.
 - d) Acute bronchopneumonia.
 - e) Acute tonsillitis.
 - f) Bacteraemia, septicaemia, pyaemia.

What is double quotidian fever (camel hump fever) ?

It is the double fever spike in a single day and is seen in :

1. Kala-azar (classical example).
2. Gonococcal perihepatitis or endocarditis.
3. Sometimes in acute bacterial endocarditis.

Appearance of rash in a febrile patient :

Remember the mnemonics : 'very sick person must take double tea' for appearance of rash in exanthematous fever.

- 1st day — Varicella, i.e., chickenpox (mainly in trunk; all forms seen at a time; no umbilication of vesicle).
- 2nd day — Scarlet fever (over chest, neck, scapula; mainly macular).
- 3rd day — Pox (smallpox); not seen now-a-days (peripheral distribution; rashes come in a sequence; umbilication of vesicle).
- 4th day — Measles (maculo-papular; over forehead, hairline near ears, face and trunk).
- 5th day — Typhus (macular; over shoulders, chest, extremities, palms and soles).
- 6th day — Dengue (morbilliform; over dorsum of hands and feet, trunk).
- 7th day — Typhoid or enteric fever (rose spots over abdomen, flanks and back; pale-pink; fades on pressure).

- * Never forget 'drug rash' which may appear any time or any day during fever.

Features of enteric fever in its first week :

1. Step-ladder pattern of pyrexia.
2. Frontal headache.
3. Constipation (diarrhoea, vomiting and **pafh** abdomen may be seen in children).
4. Anorexia, nausea, cough and epistaxis.
5. Flushed face with toxic look.
6. Angry looking tongue—Central coating with red tip and margins.
7. Caecal gurgling (due to presence of fluid faeces and air).
8. Relative bradycardia and rarely dicrotic pulse.
9. Rose spots appearing on the 7th day (usually appears on 7th-10th day).
10. There may be just palpable, soft and tender spleen at the end of first week.

What is typhoid state ?

Usually the untreated enteric fever patient enters into this toxæmic stage at the onset of 3rd week of illness and is manifested by some neurological (encephalitis-like) features like :

- a) Semiconsciousness or unconsciousness.
- b) Low muttering delirium.
- c) Coma vigil — Patient lies with half-open eyes but ignorant of his surroundings (staring stupor).
- d) Subsultus tendinum — Involuntary movements of fingers and wrists due to muscular twitching.
- e) Carphology — Agitated plucking of the bed sheets, and
- f) Convulsions rarely.

What is pulse-respiration ratio ?

It is seen that with per degree (°F) rise of temperature, there is an increase in respiratory rate by 2 to 3/minute. The pulse-respiration ratio goes like this :

- a) Normal pulse : respiration = 4:1 (72 : 18)
- b) Increased pulse : respiration e.g., 12 : 1 (72 : 6) is seen in narcotic poisoning.
- c) Decreased pulse : respiration e.g., 2 : 1 (112 : 56) is seen in acute lobar pneumonia.

Herpes labialis (fever blisters' or 'cold sore') with pyrexia :

These are painful and tender vesicles on the outer surface of lips, and is commonly observed in :

1. Acute lobar pneumonia (often gives clue to the side affected and stages of pneumonia).
2. Influenza.
3. Malaria.
4. Meningococcal meningitis.
5. Weil's disease.
6. Mycoplasma pneumoniae infection.
7. AIDS.
8. Physiological : sunlight, menstruation.

N.B. : Herpes labialis is due to herpes simplex virus, type 1 infection.

Fever with patch or membrane seen in the throat :

- | | |
|----------------------------------|-----------------------------------|
| 1. Acute follicular tonsillitis. | 5- Agranulocytosis. |
| 2. Faucal diphtheria. | 6- Vincent's angina. |
| 3. Infectious mononucleosis. | 7. Acute lymphoblastic leukaemia. |
| 4. Candidiasis. | |

What are the 'benefits' of fever ?

There is release of endogenous pyrogen with rise of temperature and this leads to activation of T-lymphocytes and thus, it enhances the host defence mechanism. Fever is sometimes beneficial in,

- | | |
|--|-------------------------------|
| 1. Neurosyphilis. | 3. Widespread carcinomatosis. |
| 2. Chronic arthritis (rheumatoid arthritis). | 4. Uveitis. |

Physiologic events on body temperature :

- (A) Diminished : on rising in the morning (lowest), old age.
- (B) Increased : exercise, ovulation and smoking.

Agents causing 'drug fever' :

- | | |
|-----------------------|--|
| 1. Sulphonamides. | 6 Salicylates. |
| 2. Iodides, bromides. | 7- Rifampicin ('flu syndrome'). |
| acils. | g Phenolphthalein (used in laxatives). |
| 4. Barbiturates. | 9 Quinidine. |
| 5. Penicillin. | 10 Phenytoin. |

The underlying mechanism is hypersensitivity and 50% patients have eosinophilia (remember the usual response in fever is eosinopenia). remember the

What is factitious fever ?

Patients Purposefully show false elevations of temperature which are often self-inflicted. **J** ³re usually young women. often attached to health professionals. They may infect themselves with bacteria, ingest thyroxine or falsely register higher temperature (after immersing the thermometer in hot water). The diagnosis is done by,

1. Dissociation between pulse and temperature.
2. Excessively high temperature (106°F).
3. Absence of chills, sweats, tachycardia, tachypnoea.
4. Absence of normal diurnal variation in temperature.

What is febrile convulsions ?

Young children often develop seizures with febrile illness and these are short, generalised tonic-clonic convulsions. The characteristics are :

1. Affects 2-5% of the population of young children.
2. Age range is from 6 months to 5 years.
3. Often there is a positive family history.
4. Convulsions are not related to the degree of elevation of temperature as they may occur even with moderate fever.
5. Seizures last for less than 5 minutes and is generalised.
6. There is no recurrence in majority.
7. It is not associated with interictal EEG abnormalities or any neurodeficit.

Management is done by tepid sponge bathing, antipyretics and inj. diazepam (0.2-0.5 mg/kg ^{ner} dose slow push I.V). Predictive factors for recurrence are : young age of onset, low fever, family H/O febrile convulsions and short duration of fever before convulsions.

Epilepsy may develop in less than 3% of all children with febrile convulsions. Predictive factors for future development of epilepsy are : partial onset, prolonged seizure (>10 min), more than one seizure in 2,4 hours and family H/O epilepsy.

Why there is a kink in the clinical thermometer ?

It prevents the return of the mercury column when the thermometer is taken out of the body.

Why the thermometer is triangular in cross-section ?

It magnifies the mercury-line into a wider strip to help in easy reading.

N.B.: One should always try to record oral temperature in the long and short cases.

Case 42

OEDEMA

What is oedema ?

Accumulation of excessive amount of tissue fluid in the subcutaneous tissue (or serous sacs) due to increase in extravascular (interstitial) component of the extracellular fluid volume resulting in swelling of tissue. A weight gain of several kilograms commonly precedes overt manifestation of oedema.

N B. : Total body water (60% of body weight in kg) is distributed as 2/3 ICF and 1 / 3 ECF; of the ECF, the distribution is 3 / 4 IF and 1 / 4 plasma. So in a person weighing 70 kg, intracellular fluid is 28 litre, while

9.5 litre is interstitial fluid, and 4.5 litre is the blood volume.

ICF = Intracellular fluid, ECF = Extracellular fluid, IF = Interstitial fluid.

How to demonstrate oedema clinically ?

1 First inspect the legs for any swelling.

2' Apply firm pressure for few seconds (at least 15 seconds to maximally upto 30 seconds sequentially over the medial malleolus, above the medial malleolus, lower end of tibia (medial surface) and the upper part of shin bone by the tip of right thumb, i.e., press over a bony background. Now inspect and palpate the area for any dimple or pitting. If it is seen that the dorsum of the foot is swollen, now one should press there. Do the same manoeuvre on the opposite side.

In oedema-forming states, **ambulatory patients** develop oedema on the dependent parts i.e., the ankle. 'Pitting' (i.e., the skin is indented) is the cardinal sign of subcutaneous oedema, which gradually fills in by redistribution of the displaced fluid.

3 Now examine the 'BACK', i.e., turn the patient into prone position and press the tip of right

thumb over sacrum. Sacral oedema is found in **patients confined to bed** for prolonged period. Sacrum *must be examined in all patients with oedema.*

4 Then examine for 'parietal oedema' - Press the chest piece of stethoscope or the tip of the five fingers of right hand, over the abdominal parietis or thigh for few seconds. Look for pitting oedema there. Oedema of the parietis (e.g., abdominal wall) may also be assessed by pinching the skin with thumb and index finger for few seconds (at least 5 seconds).

N B • Observe carefully for puffy face, puffy lower eyelids and scrotal oedema. It is said that oedema is also recognised by pallid and glossy appearance of the skin with less prominent superficial veins and at times by its doughy feel. Oedema* may be seen in the upper extremity, over sternum, vertebra and

forehead in a case of anasarca.

* If *upper part of body is involved* press over wrist, lateral epicondyle at elbow, sternum and forehead for demonstration of oedema.

A

How to classify oedema clinically ?

(A) **Whether it pits on pressure or not :**

I. PITTING OEDEMA—commonly seen in :

- a) Congestive cardiac failure.
- b) Cirrhosis of liver.
- c) Nephrotic syndrome.
- d) Hypoproteinaemia with severe anaemia e.g., in protein-losing enteropathy, starvation, kwashiorkor (nutritional oedema).
- e) Pericardial effusion.
- f) Constrictive pericarditis.
- g) Drugs - Amlodipine (i.e., vasodilators), liquorice, corticosteroid, oestrogens.
- h) Venous obstruction.
- i) Beriberi, epidemic dropsy.

II. NON-PITTING OEDEMA or SOLID OEDEMA—commonly seen in :

- a) Myxoedema (usually it is infiltrative oedema due to deposition of mucinous material)
- b) Lymphatic oedema (late stage) e.g., filariasis (due to organisation of protein-rich fluid).
- c) Angioneurotic oedema.
- d) Scleredema (post-streptococcal, self limited, painless oedematous induration) or initial stage of scleroderma.

(FU According to distribution of oedema :

I GENERALISED— it is known as '**anasarca**' (previously known as 'dropsy') and the causes are the same as a) b) c) and d) of pitting oedema. Points e) and f) too may develop into anasarca.

II LOCALISED— the common causes are,

- a) Venous obstruction- Pregnancy, SVC syndrome, IVC syndrome, varicose veins in legs, prolonged recumbency, deep venous thrombosis (DVT), immobilised or paralysed limb.
- b) Lymphatic obstruction (lymphoedema)- Filariasis, tuberculosis, carcinoma of the breast following radical mastectomy, radiation injury, Milroy's disease (congenital hypoplasia of lymph vessels).

c) Allergic cause— Angioneurotic oedema, acute anaphylaxis.

d) *Innate*— Caused by insect bite, snake bite, trauma, ischaemia or infections, and is probably due to liberation of histamine and other factors (increases capillary permeability).
Miscellaneous— Bruises, sprains, cellulitis, fracture, gout.

Localised causes may present as **unilateral oedema**; cyclical oedema may be unilateral.

*** Oedema may be restricted to limited region like pedal/sacral/facial or generalised (anasarca).

In the early stage, lymphatic oedema is soft and pits easily on pressure

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What is angioneurotic oedema ?

This is the swelling of dermis and subcutaneous tissue, and is of two types ; hereditary and acquired.

- It is a specific form of allergic oedema which results from hypersensitivity.
- 2. It is a solid oedema; involves deeper layer of skin; usually lasts for a few hours
- 3. Commonly localised in tongue, lips, face, eyelids etc.
- 4. The swelling develops rapidly and is pale or faintly pink in colour.
- 5. Associated with itching; may be ACE-inhibitor induced (acquired).

6. Congenital (hereditary) variety is due to C, esterase inhibitor deficiency.

- 7. May threaten life by suffocation, if larynx or tongue is affected.

How much body weight is to be increased for development of pitting oedema *

also ^rnThafnilrn^" ^ * necessary ^ demonstration of pitting oedema. It is so seen that pitting oedema occurs when the circumference of the limb is increased by 10%.

Why lymphatic oedema is intractable ?

s.itJiS,,fdTZ'cdre"TFOn of ,J,Tha"C "oW r"ULTS
of retained M "

Protein concentration in the Inter-
"pUlates, , situation which severely impedes removal.

Most reliable index of progress to treatment in a case of oedema :

Day to day alteration in body weight.

How cardiac oedema differs from renal oedema ?

(A) **Congestive cardiac failure :**

1. Patients will be dyspnoeic.
2. Engorged and pulsatile neck vein with soft, tender, mild hepatomegaly.
3. Sometimes associated with right ventricular gallop rhythm.
4. Evidence of heart disease, i.e., cardiomegaly may be present.
5. Oedema **starts in the dependent part (around the ankles and pretibial region)** and

then ascends progressively; pitting oedema; it is also present over the sacrum in a patient

coTM UehTerl 'te eveM,,g?ed'ma "

^ 'nCl of day'S "rk |sl"" b' _

(B) **Nephrotic syndrome :**

1. A history of previous renal disease may or may not be elicited.
2. **Oedema starts in the face (mainly periorbital), arms and then descends** Oedema is

t f ^ ^ Carly mOming_ Swelling of scrotal sacs and lower eyelids (due to low tissue tension, are classical; pitting oedema; sacral oedema in non-ambulatory patients)

Periorbital andsCcrSot ,TaSSIVE

SCVere hyp°Proteinaemia and hypercholesterolaemia.

In C^F oeriem H T SUBCUtaneous tissue fa— accumulation of oedema fluid there,
n CRF, oedema may be either due to renal insufficiency or cardiac failure secondary to hypertension.

Evolution of oedema in different systemic diseases :

1 ■ Congestive cardiac failure : Legs ^ face & ascites.

3 CirP^r^tiC n^drOIT : (peri°r^bital Puffiness) legs ascites (with scrotal swelling)
3. Cirrhosis of liver : Ascites ^ legs -> face.

4. Nutritional oedema : Oedema feet with puffiness of face -> ascites.

Common causes of pedal oedema :

1. Congestive cardiac failure.
2. Nutritional oedema, protein-losing enteropathy, hypoalbuminaemia due to any cause.
3. Cirrhosis of liver.
4. Varicose veins, deep vein thrombosis; abdominal mass (tumour, cyst, pregnancy).
5. Filariasis or chronic lymphatic obstruction, Milroy's disease.
6. Nephrotic syndrome.
7. Cyclical oedema (female > male; temporal relationship with menstrual cycle).
8. Drug-induced - Nifedipine or amlodipine, fludrocortisone, NSAIDs, oestrogens, corticosteroids.
9. Miscellaneous— Pericardial effusion, constrictive pericarditis, immobility (e.g., hemiplegia), postural (inactive persons or prolonged immobilisation), cellulitis, over inflamed joint, lipaedema (abnormal fat deposition in the hips, thighs and legs sparing feet; only in women), irradiation.

* Unilateral—deep vein thrombosis, cellulitis, filariasis (early stage), hemiplegic side and trauma;
Bilateral—other causes mentioned in the question above.

** In oedema of legs, try to determine its upper level (e.g., oedema to mid-thigh etc).

Common causes of facial puffiness :

1. Familial; facial cellulitis.
2. Hypoproteinaemia (nephrotic syndrome, acute nephritis, severe anaemia, protein-energy malnutrition).
- 3 Congestive cardiac failure, pericardial effusion, constrictive pericarditis.
4. Myxoedema.
5. SVC syndrome.
6. Cushing's syndrome.
7. Angioneurotic oedema.
8. Trichinosis as a result of infected pork ingestion.

* 'Conjunctival oedema' is common in hypoalbuminaemia. Weil's disease, Graves' disease, fluid overload and SVC syndrome.

Different mechanisms of oedema formation :

1. Low plasma oncotic pressure e.g., hypoproteinaemia (in cirrhosis of liver).
2. High capillary hydrostatic pressure e.g., congestive cardiac failure, DVT.
3. Increased capillary permeability e.g., acute inflammation, amlodipine-induced.
4. Obstructed lymphatic drainage e.g., filariasis, radiation.

Why there is oedema in heart failure ?

- 1 Impairment of renal blood flow results in more salt and water resorption.
2. Increase in venous pressure (thus, in right-sided heart failure there is venous congestion in systemic circulation i.e., there is development of oedema, and in left-sided heart failure there is pulmonary venous congestion).
3. Secondary hyperaldosteronism.
4. Increased level of ADH.
5. Some lymphatic factors may play a role.
- 6 Chronic passive congestion of liver from heart diseases (produces hypoalbuminaemia).

* In 'lipodermatosclerosis' (the skin is tethered and can not expand to accommodate oedema fluid), ankle may be spared in early oedema. Usually a female patient presents with painful areas of inflammation with induration. It may produce non-pitting oedema.

** Be careful in examining a patient of anasarca as the patient may have bilateral hydrothorax, ascites, hydrocele (with or without scrotal oedema) and rarely, pericardial effusion. Oedema restricted only in the upper half of the body is commonly due to SVC syndrome.

*** **Filariasis** may be given as a spot case.

**** Examination by the **tip of the thumb** — Oedema is demonstrated. Examination by the **ball of the thumb** — a) Sternal tenderness b) Murphy's kidney punch c) Carotid massage, and d) Air sinus tenderness are demonstrated clinically.

Case 43

Clubbing

What is clubbing ?

This is the bulbous swelling of the terminal part of the fingers and the toes with an increase in the soft tissue mass, and increased antero-posterior as well as transverse diameter of the nails due to proliferation of subungual connective tissue, interstitial oedema, and dilatation of arterioles and capillaries. Clubbing is also known as Hippocratic fingers. It is a window in clinical medicine and usually a valuable clue to an underlying disease.

What is onychodermal angle ?

It is the angle formed between the nail and the nail-bed. It is also known as **Lovibond's angle**. The normal angle is approximately 160° . Clinically it is judged by the angle formed between the nail and the adjacent skin-fold. Thus, the other name for clubbing is Lovibond's sign.

How will you examine for clubbing ?

(A) First step -

i the Patient s Angers at your eye level and look tangentially. Observe the onychodermal angle. If the angle is 180° or more, it is said that clubbing is present.

(B) Second step -

Very early clubbing can be detected by increase in fluctuation of the nail-bed (due to softening of the nail-bed). To elicit 'fluctuation', the patient's finger (say the middle finger) is placed on the pulp of examiner's two thumbs and held in this position by gentle pressure with the tips of examiner's middle fingers applied on the patient's proximal interphalangeal joint. The patient's finger is now palpated over the base of the nail by the tips of examiner's index fingers. There is always some amount of fluctuation present in normal fingers. When fluctuation is obvious (due to clubbing), it gives a spongy feel and palpation of the nail-bed may give the impression that the nail is floating on its bed.

* For detection of clubbing, first examine the onychodermal angle and then for fluctuation.

Alternative method for detection of clubbing :

Few clinicians prefer this method. Place the nails of two fingers of two hands (preferably the thumbs or ring fingers of both hands are taken for examination) face to face and look for the diamond-shaped area formed between the two nails and the proximal nail-folds. The normally formed diamond-shaped area is obliterated in the presence of clubbing. This is known as *window sign* or *Schamroth's sign*.

Digital index :

It is an objective measurement (or numerical assessment) of clubbing. For individual finger, the circumference at nail-bed (i.e., at the site of onychodermal junction) is divided by the circumference at the distal interphalangeal joint. The individual digit ratio of 10 fingers are added. The final value is now divided by 10, and if it is found to be > 1 , clubbing is said to be present.

Degree of clubbing :

1° Clubbing - Increased fluctuation of the nail bed with loss of onychodermal angle.

2° Clubbing - 1° + increase in antero-posterior and transverse diameter of the nails as well as nails become smooth and glossy with loss of longitudinal ridges.

3° Clubbing - 2° + increased pulp tissue.

4° Clubbing - 3° + wrist and ankle swelling due to HOA (hypertrophic osteoarthropathy).

N.B. : 1° and 2° are designated as MILD CLUBBING, and 3° and 4° as SEVERE CLUBBING.

What is HPOA ?

It is known as hypertrophic pulmonary osteoarthropathy where there is 'subperiosteal new bone formation at the lower end of radius, ulna, tibia and fibula, and the patient complains of swelling as well as pain in the wrist (most common) and ankle; the elbow and knee joint may be affected. The joints

SZnT- and tCnder on palPatien- Ultimately other bones like ribs, clavicle, scapula may be affected. HPOA is commonly seen in bronchogenic carcinoma (usually from squamous cell variety) and that is why the term 'pulmonary' is added. It is diagnosed by X-ray; isotope bone scanning may reveal increased periosteal activity. HOA may be familial (pachydermoperiostitis) or idiopathic.

N.B. : When you examine the patient for clubbing, always look for any swelling and tenderness in the wrist or ankle. If these are present, the clubbing is of 4°.

Causes *of clubbing* :

(A) Cardiac causes -

1. Cyanotic congenital heart diseases like Fallot's tetralogy.
2. Subacute bacterial endocarditis (SBE).
3. Rarely in cardiac tumours (e.g., atrial myxoma).
4. Eisenmenger's syndrome, primary pulmonary hypertension.

(B) Lung and pleural causes -

- | | |
|----------------------------|---|
| 1. Bronchiectasis. | 6. Pleural mesothelioma. |
| 2. Lung abscess. | 7. Pulmonary arteriovenous fistula. |
| 3. Bronchogenic carcinoma. | 8. Cystic fibrosis. |
| 4. Empyema thoracis. | 9. Rarely in long-standing fibrocaceous tuberculosis. |
| 5. Fibrosing alveolitis. | |

(C) Ulcerative colitis.

(D) Biliary cirrhosis, sometimes in hepato-cellular failure.

(E) Intestinal causes -

- | | |
|----------------------------|---------------------|
| 1. Malabsorption syndrome. | 3. Polyposis coli. |
| 2. Crohn's disease. | 4. Coeliac disease. |

(F) Normal - Found normally in some people (idiopathic).

(G) Genetic - May be a familial condition.

* Remember the mnemonic 'Clubbing' to remember the causes. Majority of the causes of clubbing remain within the CVS and respiratory system. COPD does not give rise to clubbing.

** Clubbing may be resolved after removal of tumour from lung.

*** The commonest cause in young is bronchiectasis and that in elderly is bronchogenic carcinoma.

Different types of clubbing :

1. 'Drumstick' type is seen in :
 - a) Cyanotic congenital heart diseases, e.g., Fallot's tetralogy.
 - b) Bronchiectasis.
2. 'Parrot-beak' type : Commonly seen in bronchogenic carcinoma.

Painful clubbing :

1. Bronchogenic carcinoma.
2. SBE.
3. Lung abscess.

Reversible clubbing :

1. Lung abscess.
2. Empyema thoracis.

Unilateral clubbing :

1. Presubclavian coarctation of aorta (left-sided clubbing).
2. Bronchogenic carcinoma (commonly apical).
3. Cervical rib.
4. Aneurysm of subclavian or axillary artery.
5. Erythromelalgia.
6. Arteriovenous fistula of brachial vessels.

Unidigital clubbing :

1. Hereditary (if bilateral).
2. Repeated local trauma.
3. Rarely from median nerve injury or deposition of tophi (gout).
4. Sarcoidosis.

Clubbing limited to upper extremity :

Chronic obstructive phlebitis of upper extremity as a result of chronic I.V drug addiction (e.g., heroin).

Clubbing limited to lower extremity (differential clubbing) :

In infected abdominal aortic aneurysm; sometimes in PDA with reversal of shunt (with cyanosis).

What is 'acute' clubbing ?

When the clubbing develops very rapidly e.g., in lung abscess and empyema thoracis, clubbing may develop as early as 10-14 days after the onset of illness, and is known as 'acute' clubbing.

Why the pulp tissue is increased ?

This is due to,

1. Proliferation of subungual connective tissue.
2. Interstitial oedema.
3. Dilatation of arterioles and capillaries, or opening of anastomosing channels at nail-bed.

Most *reliable' early sign of clubbing :

Loss of normal onychodermal angle. One of the earliest sign is increased fluctuation of the nail-bed though not always reliable.

Causes of 'pseudoclubbing' :

This is due to subperiosteal bone resorption of terminal phalanges (there is absence of soft tissue proliferation and increased curvature of nails) and is seen in,

1. Scleroderma.
2. Acromegaly. ■
3. Hyperparathyroidism.
4. Leprosy.
5. People working with vinyl chloride (acrosteolysis).

Which finger is usually affected first in clubbing ?

Usually the index finger.

Points to note when clubbing is detected :

- | | |
|---|-------------------------------------|
| 1. Unilateral or bilateral. | 4. Presence or absence of dyspnoea. |
| 2. Painful or not. | 5. The degree of clubbing. |
| 3. Presence or absence of central cyanosis. | 6. Duration of clubbing. |

Possible mechanisms of clubbing :

The different hypothesis are—

1. Anoxic (arterial hypoxaemia) - This is the most important theory which leads to opening up of deep arteriovenous anastomotic channels of the terminal phalanges resulting in overgrowth of soft tissues; example - Fallot's tetralogy.
2. Toxic - The example is SBE.
3. Reflex (neurogenic) - Vagotomy often improves the clubbing in bronchogenic carcinoma.
4. Metabolic - The example is thyrotoxicosis.
5. Humoral - Increased growth hormone, parathormone, bradykinin, e.g., in acromegaly.
6. Pressure changes between the radial and the digital arteries.
7. Reduced ferritin (may escape oxidation in lungs and leads to dilatation of arteriovenous anastomosis by entering into systemic circulation) may play an important role (recent view).

Pathogenesis of clubbing :

1. Hereditary predisposition - An autosomal gene of variable penetrance has been detected.
2. Vasodilators, e.g., prostaglandins, bradykinins, 5-HT may be responsible for the development of clubbing. In bronchogenic carcinoma, vasodilator substances normally detoxified by lungs enter into systemic circulation unaltered.
3. *There is trapping of megakaryocytes and platelet clumps with local release of platelet-derived growth factor (PDGF) and other cytokines (TNF- α). which increase the capillary permeability and accentuate proliferation of connective tissue (latest and most acceptable theory).*

Some other uncommon causes of clubbing :

- | | |
|--|---------------------------|
| 1. Thyrotoxicosis (part of thyroid acropachy). | 4. Sarcoidosis. |
| 2. Asbestosis. | 5. Amoebic liver abscess. |
| 3. Occupational i.e., in jackhammer operators. | 6. Portal cirrhosis. |

What is koilonychia ?

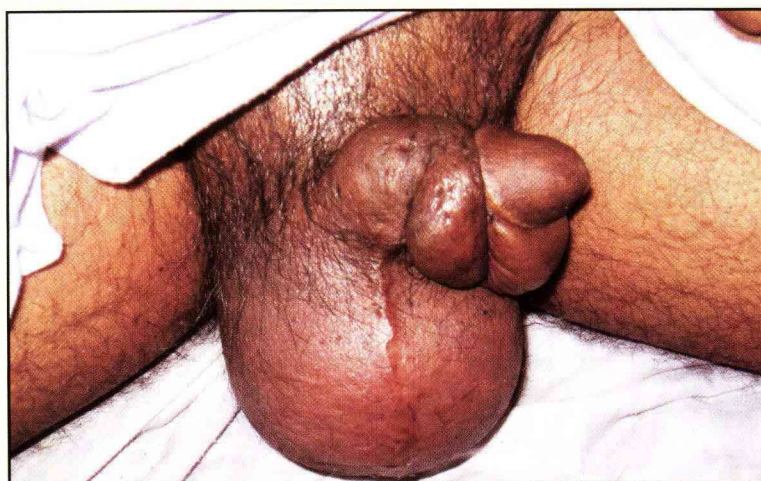
It is the 'spoon-shaped deformity' of the nail usually found in chronic iron deficiency anaemia.



Loss of pulp tissue in right middle finger and left great toe in **severe atherosclerosis**. The patient had H/O intermittent claudication



Deep venous thrombosis of calf muscles (with some eczematous changes in lower part of leg)



Oedema of the genitalia (scrotum and penis) in nephrotic syndrome



Muscle haematoma (left thigh) and monoarthritis of left knee joint in **haemophilia**



Necrobiosis lipoidica diabetorum (NLD) – yellowish plaque-like lesion which has ulcerated with atrophic centre along with surrounding spreading erythema, classically seen over the shins in diabetes mellitus

Clinical Atlas



A **cretin** aged 18 years presented with short stature, coarse facies like depressed bridge of the nose, thick lips, broad flat nose and an idiotic look, and **pot-bellied abdomen with umbilical hernia**



Achondroplasia characterized by dwarfism with short limbs and a relatively large head. Normal development of external genitalia is seen



Pituitary dwarf. Juvenile facies, thick, short and plump (chubby) with normal intelligence



Down's syndrome. A cheerful idiot



→ **Precocious puberty** in a boy aged 7 years

Koilonychia develop, as a result of retarded growth of the nail plate. Kollos' means hollow and 'onych' means nail. Toe nails are affected earlier than the finger nails.

How to **examine for koilonychia** ?

Bring the patient's fingers at your eye level and look tangentially (same as clubbing). Observe as well as palpate the nail plates for any flattening or spooning.

Stages of koilonychia :

There are three stages :

- (A) First **stage**- 'Stage of brittleness', where the nail becomes brittle and [^]ridges.
- (B) Second stage - 'Stage of flattening (platynychia). where the nail is thin, flat and without longitudinal ridges.
- (C) Third stage - 'Stage of spooning', where the nail becomes concave.

Causes of koilonychia :

1. Long-standing iron deficiency anaemia (commonest).
2. Idiopathic or familial.
3. Onycholysis due to any cause may give rise to spooning of nails.
4. Overuse of solvent (e.g., nail varnish remover) or detergents.

N.B. ⁵ After examining for clubbing, examine the other hand and next examine the toes.

Case 44

MOON FACE

Describe the face :

The face looks bloated and rounded (like the full moon).

Common causes of moon face :

1. Cushing's syndrome (and Cushing's disease)-with hirsutism and facial plethora.
2. Nephrotic syndrome (marked periorbital oedema; with baggy lower eyelids)
3. Acute glomerulonephritis (periorbital, oedema with narrowing of palpebral fissure,).
4. Prolonged steroid therapy (often with hirsutism and facial plethora).
5. Myxoedema (with baggy lower eyelids).
6. Superior mediastinal syndrome (with prominent veins in forehead and temple region).
7. Angioneurotic oedema of face (lips may be swollen; patchy involvement).
8. Subcutaneous emphysema of face, extending from chest (asymmetrical swelling).

« **'Periorbital oedema (or puffiness)**» is characteristic[^]

It usually

favours accumulation of fluid around the eyes.

** Read individual section for description.

'Check list' prior to steroid therapy :

1. H/O diabetes mellitus.
2. Pyogenic infection anywhere in the body.
3. Presence of active tuberculosis.
4. H/O peptic ulcer, gastritis or positive occult blood in stool.
5. Uncontrolled hypertension.
6. Evidence of osteoporosis.
7. Bleeding tendencies.
8. Previous H/O any psychological disorder.

Common indications for steroid therapy :

1. Addison's disease; Sheehan's syndrome.
2. Anaphylactic shock.
3. Acute severe asthma.
4. Circulatory shock.
5. Acute lymphoblastic leukaemia.
6. SLE and other collagen diseases.
7. Rheumatoid arthritis.
8. Nephrotic syndrome.
9. Acute rheumatic carditis.
10. Increased intracranial tension.
11. Interstitial lung disease.
12. In organ transplants to prevent rejection.
13. Lymphoma.
14. Cranial arteritis.
15. Vasculitides.
16. Ulcerative colitis.

How to reduce the side effects of corticosteroid ?

Give only in established indications with lowest possible dose, single dose, alternate days therapy than daily dosing, morning dose with minimal possible duration; curtail sodium and calorie intake, supplement with calcium and/or vitamin D, and bisphosphonates, H₂RA/PPI—may minimize the side effects on prolonged use.

Side effects of prolonged steroid therapy :

(A) Metabolic effects— More or less identical to those seen in Cushing's syndrome e.g., hyperglycaemia, hypokalaemia, hypochloraemia, alkalosis, sodium and fluid retention, osteoporosis etc.

(B) Suppression of hypothalamo-pituitary-adrenal (HPA) axis — In a patient with suppressed HPA axis, it may take months or years to recover fully. The dosage of steroid given only in the morning, or on alternate days, or gradual withdrawal will produce less suppression of HPA axis. Treatment for 3 weeks or less with prednisolone < 5 mg/day will not suppress adrenal axis significantly.

(C) Features like Cushing's syndrome— Moon face, truncal obesity, growth retardation, acne, hirsutism, purple striae, easy bruising, osteoporosis, avascular necrosis of bone, peptic ulcer, pancreatitis, hypertension, diabetes mellitus, glaucoma, cataract, CCF, psychosis, myopathy, impotence.

* Morning cortisol level in serum is low in iatrogenic Cushing's syndrome in comparison to true Cushing's syndrome where the level is high.

Differences between ACTH and corticosteroid therapy :

	ACTH	Corticosteroid
1. Administration	Parenteral	Oral and parenteral
2. Sodium retention	More marked	Less marked
3. Adrenal androgen	Increased	No rise
4. Adrenal atrophy	Absent	Present
5. Acne	More	Less than ACTH

Effects of sudden withdrawal of steroid therapy :

The patients should be advised to avoid sudden drug withdrawal as it may produce :

1. Suppression of HPA axis.
2. Acute adrenal insufficiency.
3. Pseudotumour cerebri (specially affecting young and obese women; there is headache, papilloedema, constricted visual field with absent neurological signs, dilated ventricles on CT scan; idiopathic condition; may be treated with steroid or ventriculo-peritoneal shunt).

What is 'facies' ?

It is the 'expressions in the face' of the patient, and very often these facial signs collectively lead to prompt aetiological diagnosis of a disease. The facial appearance may be anxious, sad, calm, happy, cachectic, pyrexial, toxic, dehydrated, puffy or may show some specific characteristic of a particular disease, e.g., parkinsonism, thyrotoxicosis or acromegaly. As face is known to be 'mirror of mind', reading of face is very important in clinical medicine.

Identification points (moon face) :

- (A) Rounded and bloated face — Moon face.
- (B) With obesity — Cushing's syndrome, prolonged steroid therapy, myxoedema.
- (C) With engorged neck veins — SVC syndrome, acute glomerulonephritis.
- (D) Expressionless face — Myxoedema.
- (E) Malar flush or plethora of face — Myxoedema or Cushing's syndrome.

- (F) With hirsutism — Cushing's syndrome or prolonged steroid therapy.
- (G) With breathlessness — SVC syndrome or receiving steroid for bronchial asthma.
- (H) With anasarca — Nephrotic syndrome.
- (I) With pallor — Nephrotic syndrome, myxoedema, SVC syndrome.

* If after careful and meticulous **examination of face** (in any patient), no diagnosis (of any 'specific facies') is reached, always try to look for :

- 1) Tongue (cyanosis, severe anaemia), 2) Pseudoptosis or partial ptosis, 3) Mild exophthalmos or enophthalmos, 4) Malar flush, 5) Masked facies, 6) goitre (though not a structure within the face), and
- 7) Ask the patient to show the upper teeth (facial palsy).

Case 45

DWARFISM (CRETINISM)

What is dwarfism ?

Dwarfism or short stature is a condition where the height of a person is much below in relation to chronological age and sex of that person as compared to a normal individual.

Common causes of dwarfism or 'short stature' :

1. Heredofamilial.
2. Constitutional (delayed growth and puberty).
3. Protein-energy malnutrition e.g., marasmus and kwashiorkor (**commonest cause of dwarfism in developing countries**).
4. Rickets.
5. Cretinism.
6. Pituitary dwarf.
7. Achondroplasia; mucopolysaccharidosis; progeria.
8. Cyanotic congenital heart diseases e.g., Fallot's tetralogy.
9. Gross kyphoscoliosis.
10. Down's syndrome, Turner's syndrome, Cushing's syndrome, Noonan's syndrome.
11. Pseudohypoparathyroidism.
12. Cystic fibrosis, bronchiectasis, thalassaemia major, coeliac disease, rheumatoid arthritis from childhood, uncontrolled type 1 diabetes mellitus, intrauterine growth retardation.

What is cretinism ?

Cretinism—hypothyroidism since birth due to partial or complete failure of thyroid gland (commonly due to thyroidal agenesis). Juvenile myxoedema — onset of hypothyroidism since childhood but develops before puberty (commonly due to dys hormonogenesis); myxoedema — onset of hypothyroidism after puberty (probably Hashimoto's thyroiditis is the commonest cause).

Describe the facies in cretinism :

The facies resembles an old man's face :

1. Appearance is dull, expressionless and idiotic; large head with sparse scalp hair.
2. Depressed bridge of the nose, broad flat nose with big nostrils.
3. Hypertelorism (widely set eyes) with wrinkling of eyebrows; narrow palpebral fissures.
4. Sparse, coarse and brittle hair with dry skin.
5. Lips are thick and everted with big, fissured, protruded tongue (macroglossia).
6. Delayed dentition with hoarse voice.

Other morphologic characteristics of cretin :

1. Short stature but maintains the infantile proportion, i.e., upper segment > lower segment.
2. The patient is lethargic and apathetic.
3. Markedly retarded intelligence. Memory is grossly impaired.

4. Skin is dry, scaly, rough and cold; skin looks pale yellow (i.e., carotenaemia).
5. Thick and short neck with presence of supraclavicular pad of fat.
6. Pot-bellied abdomen with umbilical hernia.
7. Flaccid muscles with marked hypotonia.

Features of hypothyroidism in neonatal period :

1. Prolonged physiological jaundice, 4. Feeding problem (difficulty in sucking),
2. Hoarse cry, 5. Constipation, and
3. Somnolence, 6. Delayed milestones of development (in later months).

How to differentiate between cretin and pituitary dwarf?

The characteristics of cretin (thyroid dwarf) are :

- a) Depressed bridge of the nose; coarse facies.
- b) Lack of intelligence, and
- c) More or less, normal genital development.

The characteristics of pituitary dwarf are :

- a) 'Juvenile' facies; characteristically 'chubby'.
- b) Normal intelligence, and
- c) Sexual infantilism.

Laboratory diagnosis of cretinism :

A) BLOOD -

- a) High serum cholesterol.
- b) Low radioactive I^{131} uptake.
- c) T_4 level is low with high TSH level.

* In neonates, heel-prick blood sample is used.

B) ECG - Low voltage.

C) SKELETAL X-RAY (OF LONG BONES AND PELVIS) -

- a) Delayed closure of epiphysis (i.e., bone age is less than that of chronological age).
- b) Delayed dentition.
- c) Epiphyseal dysgenesis - Instead of one epiphysis, there are multiple epiphysis (DIAGNOSTIC).

Treatment of cretin :

1. Very small dosage of L-thyroxine is needed. Initial dose is 10 mg to 25 ng, and the dose is adjusted according to clinical improvement and biochemical findings.
2. Rehabilitation.

Differentiation of dwarfism by body ratio :

(A) Upper segment = Lower segment is seen in :

- a) Heredofamilial,
- b) Constitutional, and
- c) Pituitary dwarf.

(B) Upper segment > Lower segment is seen in :

- a) Achondroplasia,
- b) Cretinism.
- c) Juvenile myxoedema.

(C) Upper segment < Lower segment is seen in :

— Spinal deformities.

* Remember, short stature may be associated with features of delayed puberty.

Characteristics of Turner's syndrome (gonadal dysgenesis) :

The genetic constitution is 45, XO; usually recognised at puberty in girls ; short stature, webbed neck, low hair line, 'shield-like' chest, widely spaced nipples, cubitus valgus and mild mental retardation. Primary amenorrhoea, failure of development of secondary sexual character, streak gonads, sterility, peripheral lymphoedema, short 4th metacarpals and coarctation of aorta are associated features.

What is achondroplasia (achondrodysplasia) ?

Dwarfism in achondroplasia results from decrease in the proliferation of cartilage present in growth plate. It is an autosomal dominant disease and is usually recognised at birth. The dwarf jokers we see in the circus are usually achondroplasia patients. The characteristics are,

- a) Dwarfism.
- b) Normal mental (intelligence), dental, endocrine and sexual development; normal trunk.
- c) Short limbs.
- d) Large head with saddle nose.
- e) Lumbar lordosis; kyphoscoliosis.

What is pseudohypoparathyroidism ?

In this condition, the secretion of parathormone (PTH) is increased but due to end organ resistance, there is no response to PTH hormone and thereby, resulting in clinical features of hypoparathyroidism. It is a hereditary disorder with distinctive skeletal and developmental defects. These patients have elevated levels of serum PTH. The common features are :

- a) Short height with stocky built.
- b) Round face with short neck.
- c) Mental retardation.
- d) Epileptic convulsions.
- e) Short 4th and 5th metacarpals and metatarsals.
- f) Basal ganglia calcification.

What is progeria ?

It is the 'premature aging' mimicking senile appearance of a person. It is manifested by accelerated atherosclerosis, atrophy of subcutaneous fat, alopecia, dwarfism, mild mental retardation and skeletal hypoplasia, and is followed by angina, myocardial infarction and systemic hypertension. Death usually occurs before the age 20.

Basic investigations performed in dwarfism :

- [1. History—natal (prenatal, intranatal and postnatal) history, family and personal history].
2. Thyroid function tests.
3. Growth hormone assay.
4. Assessment of bone age (non-dominant hand and wrist are used for radiology).
5. Chromosomal karyotyping in females (e.g., Turner's syndrome).

Identification points :

Cretins are diagnosed by typical 'coarse facies' and other morphological features (there is marked physical and mental retardation; sometimes, its D/D is mucopolysaccharidosis, i.e, Hurler or Hunter syndrome). Cretins look younger to their chronological age.

If the height of an adult is < 4 feet, he/she may be called a dwarf.

Case 46**TALL STATURE****Common causes of tall stature :**

1. Constitutional (idiopathic)—the commonest cause.
2. Racial, or familial.
3. Marfan's syndrome.
4. Gigantism (cerebral) and acromegaly.
5. Hypogonadism.
6. Klinefelter's syndrome.
7. Homocystinuria (inherited disorder of amino acid metabolism)
8. Supermales (XYY) and superfemales (XXX).

What is 'build' or 'built' ?

It is the 'skeletal framework' of a person in relation to his/her age and sex as compared to a normal person. Build is classified into two groups ;

1. Short stature, and
2. Tall stature (less common than short stature).

* Short stature : Below 3rd percentile for height of normal population of same age and sex.

Tall stature : Above 97th percentile for height of normal population of same age and sex.

** **Stature** is the total height measured from crown to heel. 'Build' is important in clinical practice due to the fact that some diseases are prevalent in tall stature while some are in short stature.

*** Body constitution (somatotypes) differs from person to person. They are divided into :

- a) Normosthenic—with normal body build.
- b) Asthenic—lean bodies, narrow shoulders, thin limbs and flat chest. They are prone to viscerop-tosis and neurasthenia.
- c) Sthenic—thick trunk, short neck, broad shoulders, and thick and short limbs. They are more likely to develop hypertension and ischaemic heart disease.

Differentiate tall stature by body ratio :

The skeletal height of a person is measured from crown to heel when he/she stands erect against the wall to which a vertical scale is attached.

Upper segment (crown to pubis) : lower segment (pubis to heel) ratio of our body is approximately 1.8 : 1 at birth; by the age of 10 years the ratio becomes 1 : 1 and, it is approximately 0.9 : 1 in adults because the legs grow more rapidly than the trunk. Hence,

(A) Upper segment = Lower segment is seen in :

- a) Constitutional, and
- b) Pituitary causes (i.e., hyperpituitarism).

(B) Upper segment < Lower segment is seen in :

- a) Marfan's syndrome,
- b) Klinefelter's syndrome,
- c) Hypogonadism (e.g., eunuchoidism), and
- d) Homocystinuria.

(C) Upper segment > Lower segment is seen in :

- a) Precocious puberty.
- b) Adrenal cortical tumour.

Clinical features of Marfan's syndrome :

Marfan's syndrome is characterised by triad of symptoms (A, B and C described below)

(A) Skeletal abnormalities like,

1. Crown to heel height is above average (i.e., tall stature) with thin built (asthenic).
2. Lower segment > Upper segment of body.
3. Arm-span > height.
4. Hyperextensible joints (Steinberg's sign or thumb sign — in which the thumb, when opposed across the palm inside the clenched hand, extends beyond the ulnar border of the palm).
5. Kyphoscoliosis, straight back syndrome.
6. Anterior chest wall deformity (pectus excavatum and pectus carinatum).
7. High arched palate.
8. Long slender fingers and toes (arachnodactyly).
9. Metacarpal index > 8.4 (the length of each of the last four metacarpal bones is divided by the width at its midpoint, and the values are averaged).
10. Wrist sign — in which the patient can enclose his wrist with the thumb and little finger of other hand, and the digits overlap each other by at least 1 cm.
11. Loose jointedness, pes planus or pes cavus.
12. Long and narrow facies (dolicocephalus).

(B) Subluxation or dislocation of lens downwards commonly (look for iridodonesis). Myopia, retinal detachment and blue sclera may be seen.

(C) CVS (CVS abnormalities are major cause of morbidity and mortality) -

1. Aortic incompetence.
2. Dissecting aneurysm of ascending aorta.
3. Mitral valve prolapse.

(D) Others—Spontaneous pneumothorax, cystic bronchiectasis, striae over shoulders.

* Intelligence and sexual development are normal in Marfan's syndrome.

** Bernard Jean Antonin Marfan (1858-1942) was Physician in Paris, France.

*** Lean and thin persons with long, slender fingers—loosely termed as 'marfanoid habitus'.

Where lies the defect in marfan's syndrome ?

Not truly known but defective collagen cross-linking may be responsible for the abnormalities in the supporting tissue. Recently it is said that mutation of fibrillin-1 and fibrillin-2 genes may be responsible for Marfan's syndrome. It is an autosomal dominant disease.

Arm-span versus height :

Normally, the arm-span (i.e., distance between fingertips when hands are stretched parallel to the ground) is equal to the height of a person. Arm-span becomes greater than height in conditions where lower segment > upper segment of the body (see above). Height is greater than arm span in hyperpituitarism, precocious puberty and adrenal cortical tumour.

Diagnostic points in Klinefelter's syndrome :

1. Tall stature (due to hypogonadism) with eunuchoid body proportions in a male; obese.
2. Lower segment > upper segment of the body. Crown to pubis length < pubis to heel.
3. Gynaecomastia with loss of secondary sexual characters.
4. Phenotypically male with small, firm testis; small penis; male psychosexual orientation.
5. Mental subnormality (mild).
6. Azoospermia (sterility) with elevated levels of plasma and urinary gonadotropins.
7. Chromation (Barr) body is usually present (chromosomal pattern is 47, XXY) in buccal smear preparation.

* Eunuchoidism, gynaecomastia and hypogonadism are triad for Klinefelter's syndrome.

Features of homocystinuria :

1. It is due to reduced activity of the enzyme cystathionine (3-synthase).
2. Mental retardation; autosomal recessive inheritance.
3. Displacement of lens downwards.
4. Life-threatening thrombotic episodes may occur.
5. Elevated plasma methionine and homocysteine levels.

What is meant by eunuchoidism ?

In a general sense, androgen loss reflects eunuchoidism. If testicular failure occurs before puberty it results in hypogonadism, lack of sexual maturation and tall stature—collectively known as eunuchoidism.

Why there is tall stature in hypogonadism ?

Testosterone accelerates epiphyseal ossification and fusion. When there is lack of testosterone in human body, there is uninhibited longitudinal growth.

Clinical associations in 'tall stature' :

Accident-proneness, isolated signs of virilism, signs of early puberty and rarely, symptoms of hyperthyroidism.

Conclusion :

One should always examine the testes, palate (high arched), spine and CVS (for AI) in tall patients.

Case 47**EMACIATION****What is emaciation ?**

Generalised undernutrition, i.e., loss of total body fat and diminution of muscle bulk.

How do you assess nutrition at the bedside ?

Nutrition is the 'nourishment' of the body. It is clinically assessed by (semi-quantitatively) :

1. SUBCUTANEOUS FAT—assessed by picking up skinfold between the index finger and thumb. Actually, it should be measured by a special calliper (Harpender or Schofield's calliper).
2. BULK OF MUSCLE MASS—measure the mid-arm circumference or mid-thigh circumference by a tape from the elbow joint or knee joint respectively (i.e., from a fixed point).

3. **SIGNS OF VITAMINS, MINERALS AND NUTRIENTS DEFICIENCY**—assessed by the presence of glossitis, cheilosis, angular stomatitis, xerophthalmia, Bitot's spot, koilonychia etc.

Remember, even the presence of mild oedema (as a result of hypoproteinaemia), anaemia, or koilonychia indicates malnutrition. For the assessment of subcutaneous fat and muscle bulk, always compare with the other side of body to diagnose localised or generalised nature of the disease process.

* In the presence of anasarca, nutrition can not be properly assessed. Skinfold thickness in the mid-triceps region (midway between acromion and olecranon process, when the arms are hanging by the side of the body) is the most commonly used site; other sites assessed are biceps, infrascapular and supra-iliac region.

** To assess the nutritional state, clinical history including dietary history and physical examination including anthropometric measurement are necessary.

*** Average adult triceps skinfold thickness : Males 12.5 mm, females 16.5 mm.

Average adult left mid-arm circumference ; Males 25.5 cm, females 23 cm.

How the 'nourishment' of the body is divided ?

- Normal
- Underweight (e.g., malnutrition, emaciation and cachexia)
- Overweight
- Obesity

How emaciated patients look like ?

All skin and bone'. All the bony prominences become more prominent.

What is malnutrition ?

Undernutrition with signs of vitamin, mineral and essential amino acid deficiency is known as malnutrition. Malnutrition may be due to starvation, maldigestion and malabsorption. Remember, over-nutrition is also malnutrition. So, a complete dietary history is important.

What is cachexia ?

It is combined manifestations of anorexia, anaemia plus emaciation, i.e., a profound state of general ill-health. Decrease in muscle mass is known as 'sarcopenia'.

Common causes of emaciation :

1. Pulmonary tuberculosis (commonly encountered in India).
2. Diabetes mellitus (in spite of increased appetite; specially in type 1 DM).
3. Thyrotoxicosis (in spite of increased appetite).
4. Disseminated malignancy (commonly colon, lung, pancreas, stomach).
5. Acquired immune deficiency syndrome (slim disease).
6. Anorexia nervosa (psychogenic) - Rare.

* Other causes of 'wasting' or 'weight loss' are malabsorption, malnutrition, cirrhosis of liver, Addison's disease, motor neurone disease, Sheehan's syndrome. Unexplained and unintentional weight loss of > 3 Kg in the previous 6 months is known as 'significant' weight loss.

Localised muscle wasting is characteristic of disuse atrophy, poliomyelitis, motor neurone disease, muscular dystrophy, peripheral neuropathy, cord lesions and local nerve injuries.

Table 27 : Indicators of nutritional depletion

Anthropometric	Chemical	Others
1. % of weight loss (>10%) (sub- or undernutrition)	1. Serum albumin (< 35 g/L)	1. Skin anergy
2. Mid-arm circumference (< 23 cm in male < 22 cm in female)	2. Serum transferrin (< 2 g/L)	2. Lymphocytopenia
3. Triceps skinfold thickness (<10 mm in male <13 mm in female)		

Hb/6, serum haematinics (iron, TIBC, ferritin, B₁₂ and red cell folate) and prothrombin time are also indices of nutrition.

How to diagnose obesity clinically ?

Overweight — When the body weight is 10% excess over the upper limit of standard weight in relation to age and sex of that individual.

Obesity — When the body weight is 20% more than the upper limit of standard weight in relation to age and sex of that individual. 'Morbid obesity' is 100% more overweight than the upper limit of standard weight.

Clinical assessment —

1. Body weight > 20% above the ideal body weight or BMI > 30 (see below).
2. When measured by a special calliper, a fatfold thickness of greater than one inch in the inferior angle of scapula (male) or the mid-triceps region (female) is known as obesity.
3. Ponderal index < 12

$$\text{Ponderal index} = \frac{\text{Height in inches}}{\text{Weight in pounds}}$$

* Obesity can be quantified by anthropometry (i.e., skinfold thickness), densitometry (underwater weight), CT or MRI scan (measures mesenteric fat).

How do you classify obesity clinically ?

1. Android—excessive fat deposition at waist.
2. Gynoid—excessive deposition of fat at hip or thigh.
3. Truncal—excessive fat deposition on face, neck and trunk (abdomen).

How can you assess obesity by body mass index (BMI) or Quetelet index :

BMI (a measure of generalised obesity) is obtained by calculating a person's weight in kilograms and dividing it by the person's height in metres squared (kg/m^2). For example, an adult male weighing 60 kg with a height of 1.70 metres has a BMI of $60/1.70^2 = 20.7$; BMI is height-dependent and thus short and tall persons of similar proportions have a similar BMI. BMI in children is less reliable than adults as an indicator of excess body fat. WHO classification of obesity is :

Category	BMI range
Underweight	< 18.5
Healthy weight	18.5 - 24.9
Overweight	25.0 - 29.9
Moderately obese	30.0 - 34.9
Severely obese	35.0 - 39.9
Morbidly obese	> 40

Abdominal or truncal obesity can be clinically measured by '**waist-hip ratio**' at the bedside. The ratio is measured by dividing the waist circumference in centimetres by the hip circumference in centimetres. The waist is usually measured at its narrowest point whereas the hips are measured at the fullest point around the buttocks. Truncal obesity is important (specially the mesenteric fat) because it is more atherogenic than peripheral fat, and is strongly correlated with insulin resistance. In a true sense, waist circumference is a strong predictor of truncal obesity.

Waist circumference : It is measured mid-way between the superior iliac crest and the lower costal margin in the midaxillary line. Substantially increased waist measurement in males > 102 cm and in females > 88 cm indicate high-risk abdominal obesity associated with metabolic complications.

Hip circumference : It is measured at 1/3rd the distance between anterior superior iliac spine and the patella, or from greater trochanter of one side to greater trochanter of other.

Systemic hypertension, diabetes mellitus, ischaemic heart disease, hyperlipidaemia and insulin resistance may be associated with abdominal (truncal) obesity, and the morbidity and mortality risks are increased if the ratio is found to be > 1.0 in males or > 0.9 in females.

The BMI is useful in the assessment of both obesity and malnutrition.

Common causes of obesity :

1. Idiopathic (simple obesity).
2. Physical inactivity (sedentary habit).
3. Cushing's syndrome.
4. Hypothyroidism (primary).
5. Hypothalamic disorders (Froehlich's syndrome or injury).
6. Insulinoma.

7. Laurence-Moon-Biedl syndrome, Prader-Willi syndrome.
8. Iatrogenic — Prolonged therapy with corticosteroid or oestrogen.

* Causes of '**weight gain**' are,

1. Different causes of obesity, and
2. Increased fluid retention due to nephrotic syndrome, CCF, cirrhosis of liver, pregnancy.

Essential amino acids :

(A) Essential (can not be synthesised in humans but are indispensable for synthesis of some important proteins) :

- | | | |
|--------------|-----------------|-----------|
| • Tryptophan | • Threonine | • Valine |
| • Histidine | • Isoleucine | • Lysine |
| • Methionine | • Phenylalanine | • Leucine |

(B) Conditionally essential (if there is adequate dietary supply, they can be synthesised from other essential amino acids) :

- | | |
|------------|-----------|
| • Cysteine | • Proline |
| • Tyrosine | • Glycine |
| • Arginine | |

Features of hypoalbuminaemia :

(A) Causes : PEM, chronic liver disease (e.g., cirrhosis of liver), protein-losing enteropathy, nephrotic syndrome, burns, trauma with sepsis (hypercatabolic states).

(B) Normal serum level : 3.5-5.5 g/dl (usually < 2.5 g/dl have serious hypoalbuminaemic features).

(C) Features : Weakness, dependent oedema, anasarca, conjunctival oedema with oedema of eyelids, polyserositis, ascites, leuconychia and increased susceptibility to infection.

Signs of vitamin A deficiency :

1. Toad's skin - Follicular hyperkeratosis (phrenoderma).
2. Xerosis of the conjunctiva, Bitot's spot, xerophthalmia and keratomalacia.
3. Recurrent respiratory and gastrointestinal infection.

N.B. : Most important symptom is night blindness (other causes of night blindness or **nyctalopia** are retinitis pigmentosa, peripheral chorioretinitis, retinal detachment, zinc-deficiency and malingering).

Bitot's spot is whitish, raised, foamy and triangular spot with its base towards limbus; it is present on the bulbar conjunctiva, a little away from the limbus.

* **Essential fatty acid** (linoleic acid, linolenic acid and arachidonic acid) deficiency also cause phrenoderma.

** Other causes of xerophthalmia in clinical practice are Sjogren's syndrome, chronic conjunctivitis, anaesthetic cornea, Stevens-Johnson syndrome, burns and pemphigoid.

Signs of riboflavin (vitamin B₂) deficiency :

1. Magenta-coloured tongue (glossitis).
2. Cheilosis (cracks and fissures at mucocutaneous junction of the lip), angular stomatitis.
3. Keratitis, vascularisation of cornea, corneal opacity, angular blepharitis.
4. Seborrhoeic dermatitis (face, scrotum) with loss of hair; fissures in the nasal orifice.
5. Anaemia.

Signs of nicotinic acid deficiency :

1. Cheilosis, angular stomatitis.
2. Raw-beefy tongue—Red, swollen and painful tongue.
3. Dermatitis (pellagra)—Specially seen on the exposed part of the body with erythema, cracking, desquamation and exudation in acute cases. There may be roughening and pigmentation of skin. Dermatitis in the neck (Casal's collar) is pathognomonic.
4. Dementia and delirium (acute form).
5. Diarrhoea.

* Common causes of **cheilosis** are,

- a) Iron deficiency anaemia.
- b) Deficiency of riboflavin, folic acid and vitamin B₁₂.
- c) Exposure to cold weather (cracked lip—in the middle of lower lip).
- d) Nicotinic acid deficiency.

Signs of pyridoxine (vitamin B₆) deficiency :

Though vitamin B₆ deficiency is rare, it may produce :

1. Polyneuropathy, 2. Mild depression (in women taking oral contraceptive pills), 3. Hypochromic anaemia, 4. Convulsions, and 5. Cheilosis, glossitis, seborrhoea.

* Uses of pyridoxine in clinical practice are :

- a) Along with INH (to prevent peripheral neuropathy), penicillamine, L-dopa and cycloserine therapy (these drugs are pyridoxine-antagonist and may produce convulsions).
- b) Sideroblastic anaemia.
- c) Neonatal convulsions.
- d) Vomiting of pregnancy.
- e) Rare diseases like xanthurenic aciduria, cystathioninuria, oxaluria.
- f) High doses of vitamin B₆ is beneficial in carpal tunnel syndrome, diabetic neuropathy, autism, schizophrenia and premenstrual syndrome.

** Increased homocysteine level in plasma is associated with acute myocardial infarction and stroke. Folate (low dose), vitamin B₁₂ and B₆ supplementation reduce the homocysteine level in plasma efficiently.

Signs of dehydration :

1. Face is drawn (facies Hippocraticus).
2. Shrunken eyes.
3. Pinched-up nose.
4. Parched lips.
5. Hollowness of temporal fossa.
6. Depressed anterior fontanelle (infants).
7. Tongue is dry and coated.
8. Skin is dry and wrinkled; subcutaneous tissue feels lax.
9. The sign of ridge' - If the skin is pinched up by index finger and thumb, and then released, a ridge is formed which subsides slowly (in severe dehydration) instead of it springing back with normal elasticity, i.e., there is loss of elasticity of skin (i.e., loss of skin turgor).
10. The eyeballs are soft due to lowering of intraocular tension.
11. Hypotension with weak pulse.
12. Scanty, dark, concentrated urine of high specific gravity.

* Assess hydration by skin elasticity, intraocular tension and recording of BP (specially, postural hypotension).

Case 48**EXAMINATION OF NECK VEINS****Internal or external jugular vein ?**

Always examine the internal jugular vein; external jugular vein is less reliable because it is:

1. Superficial and prone to kinking.
2. Does not directly drain into SVC,
3. May have valves, may pierce fascia,
4. Engorged, if patient wears a tight collar,
5. Not a direct reflector of central venous pressure (CVP).

* Internal jugular vein is in direct continuity with the right atrium and reflects the pressure changes in the right atrium as well as CVP. As it is deeply situated, the internal jugular vein is difficult to visualise.

N.B. : Recent view is that both external and Internal jugular veins have valves and as internal jugular vein is difficult to visualise, the external jugular vein is gaining importance.

Prerequisites for accurate assessment of neck veins :

1. The patient should be reclining supine at 45° (angle formed by trunk and the level of the bed at waist is 45°).
2. Neck muscles should be 'relaxed' properly (examination should always be done with a 'back-rest' or properly adjusted pillows; if 'back-rest' is not available, patient's body can be lifted and supported on your arm to make an angle of 45° but proper relaxation of neck muscles is not achieved by this method).

3. Good light is necessary for visualisation of neck veins. A light source e.g., a torch illuminated from behind (tangential application) helps to visualise the waves in a better way.
4. It is better to observe the right internal jugular vein (the innominate vein may be compressed by the aortic knob which damps and elevates the venous pressure in the left jugular vein).
5. It is better to stand on the right side with patient's face slightly turned to the left for observation of right-sided neck vein.
6. One should look in between the two heads of sternomastoid muscle and if it seems full, observe the uppermost point of distension (one may identify the internal jugular pulsation with the help of hepato-jugular reflux, or by occluding it with gentle digital pressure above the clavicle).

N.B. : If in the sitting position neck veins are fully engorged, it is not necessary to examine the patient at 45°, and in that situation measure venous pressure in sitting position. Similarly, the patient may have to lie flat if the JVP is very low.

Table 28 : Bedside differentiation between venous and arterial pulsation in neck

Venous pulsation	Arterial pulsation
<ol style="list-style-type: none"> 1. Wavy or undulating 2. Better seen than felt 3. Becomes prominent on expiration 4. Becomes prominent on lying down 5. Becomes prominent on application of abdominal pressure (hepato-jugular reflux) 6. Prominent movement is inward 7. Can be abolished by gentle digital pressure applied above the clavicle (important point of differentiation in the examination hall) 8. Two positive waves are seen (when in sinus rhythm) 9. It has a definite upper level 10. Pulsatile displacement of the ear lobes in high CVP 	<ol style="list-style-type: none"> 1. Jerky or abrupt 2. Better felt than seen 3. No change 4. No change 5. No change 6. Prominent movement is outward 7. Does not affect arterial pulsation 8. One positive wave is seen (when in sinus rhythm) 9. No such level is seen 10. Absent in arterial pulsation

Causes of increased jugular venous pressure (JVP) :

(A) ENGORGED AND PULSATILE NECK VEIN :

1. **Congestive cardiac failure (commonest cause of raised JVP).**
2. Chronic constrictive pericarditis.
3. Cardiac tamponade.
4. Tricuspid valve disease (TI and TS).
5. Restrictive cardiomyopathy.
6. Chronic cor pulmonale.
7. Pulmonary thromboembolism.
8. After massive fluid infusion or blood transfusion, pregnancy, acute nephritis (i.e., salt and water retention).
9. Complete heart block (from time to time cannon waves are seen).

* T JVP is the first sign to appear and last sign to disappear in CCF or right heart failure.

(B) ENGORGED AND NON-PULSATILE NECK VEIN :

1. **Superior mediastinal syndrome (commonest).**
2. Rarely in chronic constrictive pericarditis.
3. Valsalva manoeuvre.

Low jugular venous pressure (JVP) :

1. Dehydration (commonest).
2. After haemorrhage (hypovolaemia) or patient in hypovolaemic shock.
3. After massive diuretic therapy.

* JVP is increased in cardiogenic shock.

Points observed in neck vein at the bedside :

(A) Engorged or not.

- (B) If engorged :
- Pressure,
 - Pulsation (waveforms), and
 - Hepato-jugular reflux.

Importance of hepato-jugular reflux :

When firm pressure is applied on any part of the abdomen for 10 seconds, specially over the right hypochondrium, there is transient rise in JVP as the hepatic venous reservoir is compressed (the patient reclining at 45°). This is hepato-jugular reflux (often called abdomino-jugular reflux).

In health, significant alteration of JVP does not occur by this manoeuvre. A positive hepato-jugular reflux is defined by increase in JVP on 10 seconds firm abdominal compression, which shows rapid fall back of venous column by 4 cm on release of compression. The commonest cause of a positive reflux is right-sided heart failure. Its importance lies in :

- To diagnose incipient (early stage) or overt right heart failure (CCF),
- To differentiate between arterial and venous pulsation, and
- To differentiate between obstructive and non-obstructive causes of engorged neck vein (negative hepato-jugular reflux is seen in SVC syndrome and Budd-Chiari syndrome).

Waves seen in neck vein :

Positive waves—a, c and v; actually, there are two positive waves (a and v) seen clinically as c-wave is small and rarely appreciated at the bedside. Negative troughs (waves)—x and y.

a-wave is absent in atrial fibrillation and v-wave (so called 'systolic wave') becomes very prominent in TI. Small x-descent in TI, and prominent x-descent in cardiac tamponade; slow y-descent in TS, and sharp y-descent is found in constrictive pericarditis.

Mechanism for production of different waves in neck vein :

'a' wave	—	Right atrial contraction. It is the most dominant wave in JVP.
'c' wave	—	Closure and bulging of tricuspid valve (according to few clinicians, it is due to transmitted carotid impulse).
'x' descent	—	Atrial relaxation with downward descent of tricuspid valve.
'v' wave	—	Passive right atrial filling during ventricular systole.
'y' descent	—	Right ventricular filling with atrial emptying.

* a-wave coincides with S_1 , v-wave with S_2 .

x-descent follows S_1 , y-descent follows S_2 .

What is giant a-wave (venous Corrigan) ?

When the right atrium contracts against outflow tract obstruction, giant a-wave is produced, e.g.,

- Tricuspid stenosis,
- Pulmonary hypertension, and
- Pulmonary stenosis.

* Exaggerated a-waves are the hallmark of right atrial hypertrophy.

Cannon wave :

Huge pathological (i.e., very large) a-wave is known as cannon wave. It is seen when the right atrium contracts against closed A-V valve (i.e., tricuspid valve). It is commonly found in.

- Regular—Nodal tachycardia, junctional rhythm.
- Irregular—Complete heart block, classic AV dissociation with ventricular tachycardia.

Kussmaul's sign :

In normal healthy persons, the neck venous pressure falls (so, the height lessens) on deep inspiration due to sucking of blood into the right atrium. Reverse happens after deep expiration.

But in patients with constrictive pericarditis, cardiac tamponade (less common), severe right-sided heart failure, right ventricular infarction (produces decreased right ventricular compliance) and restrictive cardiomyopathy, there is paradoxical rise in JVP after deep inspiration due to non-accommodation of increased venous return to right side of the heart. This is Kussmaul's sign and is also known as venous pulsus paradoxus.

Why the neck vein is examined at 45° ?

Regardless of body position, the centre of the right atrium lies 5 cm below the sternal angle (the 'reference point'). When a person is lying flat, the jugular veins are seen distended in the neck. If he/she

is at 90° (sitting), the upper level of vein is never seen normally. But if the person reclines at 45°, the upper level of vein is just visible above the clavicle or invisible. So, if we see the pulsating venous column far above the clavicle at 45°, the JVP is said to be raised. It is why the neck veins are always examined at 45°. In the other way, **the patient should lie at 45° in order to standardise reading.**

At 45°, the vertical distance between the top of the oscillating venous column and the sternal angle is the jugular venous pressure (JVP) and is normally 3-4 cm. Thus, 3 cm + 5 cm = **8 cm of blood or H₂O is the normal venous pressure, right atrial pressure, JVP or CVP.**

How venous pressure is measured, at bedside ?

It is measured by the vertical height of internal jugular vein above the sternal angle. Clinically it is done with the help of two postcards (or scales); one postcard is placed vertically over the sternal angle and the other is placed horizontally from the top of the oscillating venous column upto the first postcard when the patient is reclined at 45°. The junctional point between the two postcards is marked and the vertical distance from this point to the sternal angle is measured, e.g., if it is seen to be 6 cm in a patient of CCF, the JVP of that patient will be 5 cm + 6 cm = 11 cm of blood, i.e., JVP is raised.

Describe the neck vein in CCF :

- a) Engorged and pulsatile.
- b) JVP is raised (measure, if possible).
- c) Waves are seen.
- d) Hepato-jugular reflux is present.

* During elicitation of hepato-jugular reflux in a suspected patient of CCF, patient will C/O pain (as the liver is tender on palpation due to congestive hepatomegaly). Also look for bipedal oedema. These are indirect evidences in the examination hall.

What is Friedreich's sign ?

It is the rapid y-descent in the venous pulse seen in constrictive pericarditis and is due to rapid inflow of blood into the right ventricle as soon as the tricuspid valve opens.

Bedside diagnosis of constrictive pericarditis :

When acute fibrinous or serofibrinous pericarditis (i.e., from tuberculosis, bacterial infection or haemopericardium) heals and leads to obliteration of pericardial sac by granulation tissue, thick and fibrous pericardium encase the heart and the disorder is known as chronic constrictive pericarditis. It is diagnosed by :

1. Fatigue, dyspnoea on exertion, orthopnoea, loss of weight (symptoms).
2. Protuberant abdomen (ascites).
3. Peripheral oedema and systemic venous congestion.
4. Rapid, low volume pulse and pulsus paradoxus.
5. Elevated CVP with Friedreich's sign (sharp y-descent).
6. Kussmaul's sign.
7. Invisible or feeble apex beat with normal-sized heart; distant (or quiet) heart sounds.
8. Pericardial knock heard at lower left sternal border.
9. Congestive hepatomegaly with ascites; ascites > peripheral oedema.
10. Splenomegaly.

* The pathophysiology of clinical features in constrictive pericarditis are :

- 4. Ventricular filling (just like cardiac tamponade)—Kussmaul's sign, Friedreich's sign, pulsus paradoxus.
- Systemic venous congestion—↑ JVP, oedema, ascites and hepatomegaly.
- Pulmonary venous congestion—dyspnoea, orthopnoea (rare).
- 4. Cardiac output—fatigue, low volume pulse.

** One of the common D/D of constrictive pericarditis at the bedside is cirrhosis of liver.

*** **Pericardial knock** — This is an extra sound in early diastole and is probably due to restrictive effect of adherent pericardium on diastolic expansion of the ventricle.

Conclusion :

During examination of the neck veins in the examination hall, always ask for 'back-rest'. If not supplied, support the patient's trunk on your left arm to make an angle of 45°.

Although the neck veins are engorged and pulsatile in biventricular failure, it is primarily a sign of right ventricular failure.

Case 49

SPIDER NAEVI

What are these ?

These are telangiectases that consist of a centrally dilated arteriole from which numerous small vessels radiate resembling the legs of a spider. Usually it is a sign of hepato-cellular failure.

Common sites :

In the upper half of thorax (necklace area) and back of upper chest commonly; face, forearms and dorsum of hands. Rarely, they are seen in mucous membrane of oral cavity and nose.

Method of demonstration :

The central arteriole (central prominence) is occluded by giving pressure with a pinhead or glass slide, and the response is immediate blanching of the whole naevus. If the pressure is released, the naevus goes back to its original shape and colour, which shows filling from the centre to the periphery.

Size of spiders :

From a pinhead to 0.5 cm in diameter. When it attains large size, it may be seen or felt to pulsate.

Other names of spider naevi :

Arterial spider, spider telangiectasis, spider angioma, vascular spider.

Clinical associations :

- | | |
|---|---|
| 1. Cirrhosis of liver, specially in alcoholics. | 5. Sometimes, in thyrotoxicosis. |
| 2. Viral hepatitis (transient). | 6. In some healthy persons (2%)—< 3 in number, F > M. |
| 3. During third trimester of pregnancy. | 7. Oestrogen therapy. |
| 4. Occasionally, in children. | |

Aetiopathogenesis :

Spiders are formed due to hyperoestrogenaemia (diminished hepatic clearance of precursor androstenedione results in increased peripheral conversion to oestrogen). It is known that oestrogen has a dilating effect on spiral arteries of endometrium and such a mechanism may be responsible for the development of cutaneous spiders. Now-a-days, it is said that the ratio of (high) oestrogen and (low) testosterone level is the main factor for production of spiders.

Differential diagnosis :

1. Mosquito bite—The 'fresh' mosquito bite marks blanch like spiders. H/O using mosquito nets at night should be taken. Mosquito bite marks may be present anywhere in the body.
2. Hereditary haemorrhagic telangiectasis (HHT)— '*Telangiectasia*' are small dilated blood vessels which may be visible on the skin surface, particularly on the lips and *usually blanch on pressure*. Common sites of HHT are mucosal surfaces like tongue, lips, nose, palate, pharynx, oesophagus and stomach. It is generally connected with a single vessel. Telangiectasis may be seen in CREST syndrome.
3. Haemorrhagic spots—Petechiae and purpura never blanch on pressure.
4. Venous star—It is seen in persons with high venous pressure and never blanches on pressure. Common sites are back, legs, lower part of anterior chest wall and dorsum of foot. Here, the blood flows from the periphery to the central part in contrast to vascular spiders where blood flows from the centre to the periphery.
5. Campbell de Morgan's spots (cherry angioma)—These spots are very common in aged persons (size -1 to 2 mm in diameter), and are commonly seen in anterior abdominal wall and front of the chest. They do not blanch on pressure and are usually elevated from the skin surface.
- 6) Rose spots—Seen in enteric fever, specially over abdominal skin; blanch on pressure.

Why the spiders are mostly seen in necklace area ?

It is said that spiders are seen in the area drained by superior vena cava (above the line joining both nipples). Actually, the basic cause is not known but it is expected that exposure of the necklace area to sunlight may damage the skin in such a way that appropriate internal stimulus may provoke the development of spiders in the said area. Experiment shows that people in the nudist colony having cirrhosis of liver show spiders distributed all over the body.

Draw inference if a previously seen spider disappears :

1. The hepato-cellular failure is corrected (good prognosis), or
2. The fall of blood pressure in a case of hepato-cellular failure (commonly due to fresh haematemesis or melaena) may be responsible for the disappearance of spiders (bad prognosis).

Conclusion :

One should search spiders meticulously in its specified area of distribution. Spiders are very often missed, if not careful. Confirm it by 'blanching response' after application of pressure by a pinhead.

Case 50**PALMAR ERYTHEMA****What is this ?**

It is commonly seen that in a setting of hepato-cellular failure, the palms become bright red in colour (and also warm). Thus, other name of palmar erythema is 'liver palms'. The soles of the feet may be similarly affected.

Sites affected within the palm :

- | | |
|-------------------------|-----------------------------|
| 1. Thenar eminence, | 3. Pulp of the fingers, and |
| 2. Hypothenar eminence, | 4. Bases of the fingers. |

Method of demonstration :

Like the spider naevi, **palmar erythema blanches on pressure**. Here, a glass slide is used for application of pressure and the redness of palm reappears after release of pressure. In the absence of glass slide, press the patient's palm with your thumb.

* At times, the palm flushes synchronously with the pulse rate after being pressed by a glass slide.

Clinical associations :

- | | |
|---|---|
| 1. Cirrhosis of liver. | 5. Thyrotoxicosis. |
| 2. In alcoholics. | 6. Chronic leukaemias. |
| 3. Long-continued rheumatoid arthritis. | 7. Rarely, in some normal persons (familial). |
| 4. Pregnancy. | 8. Hyperdynamic circulation e.g., high fever. |

* Remember, *polycythemia* causes redness of the palm and sole; so, search for plethoric face, dusky-red oral mucous membrane and suffused conjunctiva.

Aetiopathogenesis :

Same as spider naevi, or a non-specific change indicative of hyperdynamic circulation.

Skin and appendicular changes in hepato-cellular failure :

1. Spider naevi.
2. Palmar erythema.
3. Bleeding manifestations— Due to coagulation disorder (hypoprothrombinaemia) and defects in the platelet (number and functional abnormalities) petechiae, purpura, ecchymosis may be seen.
4. 'Paper money skin'— It is usually seen in upper arms and also observed in the common sites for spider naevi. These are nothing but small blood vessels and resemble silk threads in United States' dollar bills.
5. White spots — Seen in arms and buttocks (demonstrated after cooling the skin). These are forerunners of a spider.
6. White nails (leuconychia) — This is due to opacity of the nail-bed. All the fingers may be affected but thumb and index fingers are specially involved. Hypoalbuminaemia is thought to be responsible for production of white nails (thus, sometimes seen in nephrotic syndrome).
7. Gynaecomastia— May be unilateral and is developed as a result of hyperoestrogenaemia. It is said that spironolactone therapy is the commonest cause of gynaecomastia in a patient of cirrhosis of liver.

8. Clubbing— Specially seen in biliary cirrhosis.
9. Jaundice—In case of severe jaundice, skin may become yellow. Jaundice gives some guide to the severity of hepato-cellular failure.
10. Diffuse pigmentation of the body, particularly over the face.
11. Dupuytren's contracture, specially in alcoholics.
- 1 2. Scanty pubic and axillary hair.
13. Xanthoma (in palmar creases) or xanthelasma (around eyes) are seen in primary biliary cirrhosis.

Case 51

EXAMINATION OF THE HANDS

Points to note in hands :

(A) Shake hands with the patient (to assess surface temperature, skin and the grip strength) may reveal :

- a) Warm and moist hand—thyrotoxicosis, type II respiratory failure, pyrexia (only warm).
- b) Cold and moist hand—anxiety neurosis; moist and doughy handshake—acromegaly; cold and clammy hand with peripheral cyanosis—shock.
- c) Cold and dry hand—myxoedema: only cold hand—low cardiac output (e.g., heart failure), hypothermia, Raynaud's phenomenon.
- d) Patient may not relax his hand after handshake—myotonia.
- e) Weak grip strength—weakness of the small muscles of hand or weakness of flexor muscles.
- f) Rough, thick palm—phrenoderma (vitamin A or essential fatty acid deficiency), arsenic poisoning, tylosis (hyperkeratosis of palm often associated with oesophageal carcinoma), psoriasis, verruca vulgaris, multiple corn or callosities.
- g) Shake hand producing pain — rheumatoid arthritis.

(B) Colour changes :

- a) Anaemia or pallor (the palmar creases become pale when the Hb level is < 7 g/dl).
- b) Jaundice—mainly the palms, dorsum rarely.
- c) Cyanosis—tips of fingers, nail-beds and palm.
- d) Palmar erythema.
- e) Janeway's lesion (non-tender maculopapular lesion present in thenar eminence in SBE).
- f) Carotenaemia (orange-yellow or lemon-yellow colour of palm).

(C) Finger deformities :

- a) Arachnodactyly (long slender, spider-like fingers)—Marfan's syndrome, hypogonadism.
- b) Polydactyly (extra finger)—Laurence-Moon-Biedl syndrome, associated with VSD.
- c) Syndactyly (joined fingers)—Poland's syndrome, underlying congenital heart disease.
- d) Clinodactyly (incurved finger)—commonly affects the little finger in Down's syndrome.
- e) Sclerodactyly (tight skin over the phalanges)—scleroderma.
- f) Broad hand with thick, large, square-tip fingers (spade-shaped hand)—acromegaly.
- g) Bradydactyly (short fingers)—Down's syndrome. Turner's syndrome, mucopolysaccharidosis.

(D) Clubbing :

(E) Wasting of the small muscles of hand : leprosy, MND, syringomyelia, rheumatoid arthritis etc.

(F) Pulsation :

- a) Capillary pulsation (see the section on 'Aortic incompetence').
- b) Digital artery pulsation (see the section on Aortic incompetence).
- c) Pulse—give the classical description (some clinicians may not like to include 'examination of pulse' under examination of the hand).

(G) Tremor:

- a) Static tremor.
- b) Action tremor.
- c) Intention tremor, and
- d) Flapping tremor

} read the section on 'Tremor'

(H) Swellings :

- a) Osier's node (tender papule mainly in the pulp of fingers)—seen in SBE.
- b) Heberden's node—hard bony nodules seen at the dorsal aspect of distal interphalangeal (DIP) joints in osteoarthritis.
- c) Bouchard's node—hard bony nodules seen at the dorsal aspect of proximal interphalangeal (PIP) joints in osteoarthritis.
- d) Rarely, rheumatoid nodules or nodule formation in leprosy.
- e) Gouty tophi—over dorsal aspect of PIP and DIP joints.
- f) Calcinosis—scleroderma.
- g) Oedema over dorsum of the hand. Ganglion, neurofibroma, lipoma are of surgical interest.

(I) Hand deformities :

- a) Spindle-shaped fingers (due to swelling of PIP joints)—rheumatoid arthritis, scleroderma.
- b) Swan neck deformity (hyperextended PIP joints and flexed DIP joints)—rheumatoid arthritis.
- c) 'Boutonniere' or button-hole deformity (flexed PIP joints and hyperextended DIP joints) — rheumatoid arthritis, and is just reverse of swan neck deformity.
- d) Volar subluxation with radial deviation at wrist and ulnar deviation of digits—rheumatoid arthritis.
- e) Z-deformity of thumb (hitch-hiker's thumb)—rheumatoid arthritis.
- f) Swelling of DIP joints only—osteoarthritis, psoriatic arthropathy or scleroderma. In psoriasis, telescoped phalanges may produce shortening of fingers with gross impairment of function.
- g) Dupuytren's contracture (flexion contracture of ring and little fingers due to thickening, fibrosis and shortening of ulnar side of palmar fascia). The causes are :
 - a) Repeated trauma to palm.
 - b) Alcoholic cirrhosis.
 - c) Phenytoin sodium therapy⁷ in epileptics.
 - d) Working with vibrating tools.
 - e) Manual workers (gardener).
 - f) Diabetes mellitus.
 - g) Peyronie's disease.
 - h) Idiopathic.
- h) Surgical deformities like club hand, Madelung's deformity, mallet finger etc.
- i) Claw hand—read the section on 'Claw hand'.
- j) Main d' accoucheur—metacarpophalangeal (MCP) joints are flexed, the PIP and DIP joints are extended with opposition of the thumb. It is commonly seen in tetany and also known as carpal spasm. The method of demonstration of carpal spasm by sphygmomanometer is known as Trousseau's sign. [There is another Trousseau's sign in internal medicine which is characterised by migratory thrombophlebitis in a patient with carcinoma of the pancreas.]
- k) Jaccoud's arthritis—rheumatic arthritis usually heals without deformity; very rarely, minor deformity of hand (marked ulnar deviation of MCP joints due to subluxation) may persist. Rarely, SLE and Sjogren's syndrome may develop into this type of deformity.
- l) Short 4th and 5th metacarpals in pseudohypoparathyroidism ('dimpling sign').
- m) Wrist drop—Radial nerve palsy.

* Armand Trousseau (1801-1867) was Physician, Hotel Dieu, Paris, France who actually died from pancreatic carcinoma.

(J) Nail changes :

- a) Pitting (thimble) nails—psoriasis, atopic eczema, alopecia areata etc.
- b) Beau's line—transverse ridges develop after recovery from any febrile illness, in zinc deficiency.
- c) Horder's line or splinter haemorrhage (linear longitudinal haemorrhage underneath the nails)—SBE, trauma, trichinosis, systemic vasculitis etc.
- d) Mees line—transverse white stria of 1-2 mm width appears above lunula (the white part at base of nail) in arsenic poisoning.
- e) Koilonychia 'spoon-shaped nail' is seen in chronic iron deficiency anaemia.
- f) Onycholysis—candidiasis, ringworm infection, psoriasis, lichen planus, trauma etc.
- g) Missing nail in nail-patella syndrome — presents with (hereditary) nephrotic syndrome.
- h) Nail-fold thrombi—scleroderma and other collagen vascular diseases (vasculitis).
- i) Green nail—severe pseudomonas infection.
- j) Blue lunula (azure arcs) CuSO₄ poisoning, Wilson's disease, zidovudine therapy, cyanosis and ochronosis.
- k) Red lunula—seen in CCF.
- l) Black nail—hair dyes, Peutz-Jeghers syndrome.

- m) Brown nail—hydroquinone-induced.
- n) Grey-black nail—melanoma.
- o) Bitten nails—persons with anxious personality.
- p) Fungal infection of nails (onychomycosis)—candidiasis, ringworm (tinea unguium),
- q) Shiny nails—indirect evidence of pruritus.
- r) Leuconychia (white nails)—seen in hypoalbuminaemic conditions like cirrhosis of liver, nephrotic syndrome, severe malnutrition, kwashiorkor and protein-losing enteropathy; also known as Terry's nail.
- s) Yellow nails in 'yellow nail syndrome'; other components of the syndrome are lymphoedema of the extremities and pleural effusion,
- t) Half-and-half nail (proximal half is white and distal half is pink) — seen in uraemia; also known as Lindsay nail.
- u) Platynychia (flat nails) — Hereditary, iron deficiency anaemia,
- v) Pterygium unguis (loss of skin fold around nail) - seen in lichen planus.
- w) Paronychia or whitlow (i.e., swollen and inflamed nail-bed)—working in wet conditions, diabetes, repeated trauma,
- x) Hypertrophy of nails—chronic fungal infection.
- y) Absence or atrophy—nail-patella syndrome, congenital ectodermal defect,
- z) Subungual fibroma—observed in tuberous sclerosis.
- a) Brittle nail—fungal infection, iron deficiency anaemia, psoriasis, peripheral vascular disease.

(K) Skin changes (mainly dorsum of the hand) :

- a) Hyperpigmentation.
- b) Hypopigmentation.
- c) Ichthyosis (fish skin).
- d) Palmar xanthoma—seen in familial hyperlipidaemia.
- e) Lilac-coloured knuckles in dermatomyositis (Gottron's sign).
- f) Petechiae, purpura, ecchymosis.
- g) Rash (from diseases or drugs).

(L) Miscellaneous :

- a) Nicotine staining in right index and middle fingers in chronic smoker.
- b) Single palmar crease (simian crease) in Down's syndrome.
- c) Involuntary movements like chorea, athetosis, minipolymyoclonus etc.
- d) Power of individual small muscles should be tested; sensory loss, if any, should be detected.
- e) Trophic ulcers—vasculitis, Raynaud's disease, neurological disorders (e.g., syringomyelia).
- f) Movement at different joints— to find out any restriction of movement.
- g) Loose-jointedness—tested by passive hyperextension of fingers, and even the hyperextended fingers may touch the forearm. It is commonly seen in osteogenesis imperfecta, Marfan's syndrome, Ehlers-Danlos syndrome and in some normal persons.
- h) Pulp atrophy, telangiectasia, digital infarction, digital ulceration, digital gangrene, triphasic colour response of Raynaud's phenomenon—in scleroderma or SLE (features of vasculitis).
- l) Test for finger-flexion jerk and Hoffman's sign (see the sections on 'Jerks and clonus'), grasp reflex, and test for myotonia (tapping the thenar eminence).

Examination of the hand in SLE and scleroderma :

Read the individual sections.

Examination of the hand in leprosy :

1. Read the skin changes in TT and LL varieties from the section on 'Leprosy'.
2. Claw hand with wasting of the small muscles of hand (commonly ulnar clawing affecting fourth and fifth fingers), and
3. Trophic changes — Dry and rough skin, brittle nails, loss of hair, digital ulceration and gangrene, tapering and conical fingertips with trophic ulcers, and loss of distal digits.

Examination of the hand in rheumatoid arthritis :

1. Spindle-shaped fingers (swelling of PIP joints, DIP joints unaffected).
2. Swelling of the wrist and different joints (MCP and PIP) in the hand— No swelling at DIP joints.

3. Swan neck deformity (described earlier)— Due to disruption of the volar plate of PIP joints, often with associated rupture of the insertion of flexor sublimis.
4. 'Boutonniere' or button-hole deformity (described earlier)— Chronic synovitis of PIP joints lead to rupture of central slip of extensor tendon (looks like gap of button hole) allowing protrusion of joint between two lateral slips of extensor tendon.
5. Volar subluxation with radial deviation at wrist and ulnar deviation of digits— Due to bowstring action of displaced extensor tendons.
6. Z-deformity of thumb— Due to muscle wasting, rupture of tendon and weakened joint capsule.
7. Opera glass hand appearance—Mutilating arthropathy.
8. Palmar erythema (seen in long-continued rheumatoid arthritis).
9. Joint movements are painful and restricted.
10. Grip-strength is poor.
11. Wasting of the small muscles of hand— Chiefly due to disuse atrophy; carpal tunnel syndrome, vasculitis, mononeuritis multiplex or rarely prolonged use of corticosteroids may be attributed.
12. Digital infarction, digital gangrene, splinter haemorrhage.
13. Nail-fold thrombi.
14. Carpal tunnel syndrome (with evidence of wasting of thenar muscles).
15. Rheumatoid nodule (rare in hands).
16. Trigger finger (flexion deformity from which the finger can be extended only by force).
17. Rupture of the extensor tendons of ring and little fingers resulting in 'dropped finger' (rare).
18. Purpuric rash (thrombocytopenia due to Felty's syndrome).
19. Raynaud's phenomenon.
20. Pallor (anaemia) in severe active deforming disease.

'Vasculitis' and its characteristic skin changes :

Vasculitis is the necrotising inflammation of blood vessels. Endothelial oedema and proliferation with haemorrhage contribute to the occlusion of the vascular lumen and subsequent ischaemic changes along with organ damage. Vasculitis is commonly due to infections, drugs (e.g., allopurinol), immunological disorders (e.g., SLE) and malignant diseases. It is classified according to the size of vessel involved (Chapel Hill Consensus Conference Classification).

Classification :

- (A) Large-vessel vasculitis : Giant cell arteritis (temporal arteritis) and Takayasu's arteritis (aortic arch syndrome or pulseless disease).
- (B) Medium-sized vessel vasculitis : Polyarteritis nodosa and Kawasaki disease (mucocutaneous lymph node syndrome).
- (C) Small-vessel vasculitis of arterioles, capillaries and venules : Wegener's granulomatosis, microscopic polyangiitis, Churg-Strauss syndrome, leucocytoclastic vasculitis (hypersensitivity vasculitis), Henoch-Schonlein purpura and essential mixed cryoglobulinaemia.

* SLE, Behcet's disease, scleroderma, polymyositis or dermatomyositis, SBE, rheumatoid arthritis and paraneoplastic syndromes are also associated with vasculitis.

** ANCA-associated vasculitis are Wegener's granulomatosis, Chmrg-Strauss syndrome and microscopic polyangiitis. ANCA stands for antineutrophil cytoplasmic antibody.

Dermatological manifestations ;

1. Large arteries - Digital gangrene.
2. Small arteries -
 - a) Palpable purpura.
 - b) Urticarial rashes (tender).
 - c) Morbilliform eruptions (look like measles rashes).
 - d) Maculopapular eruptions.
3. Arterioles and very small arteries -
 - a) Digital infarction.
 - b) Digital ulceration.
 - c) Nail-fold thrombi.
 - d) Vesicular or bullous lesion.
 - e) Splinter haemorrhage.
 - f) Raynaud's phenomenon.
 - g) Pulp atrophy.
 - h) Necrotic ulcer.

*** Vasculitis in collagen vascular diseases is responsible for these characteristic skin lesions in hands.

Case 52

WASTING OF THE SMALL MUSCLES OF HANDS

What are the small muscles of hand ?

(A) Thenar muscles -

1. Abductor pollicis brevis,
2. Flexor pollicis brevis, and
3. Opponens pollicis.

(C) Adductor pollicis.

(D) Palmaris brevis (superficial muscle of hand).

(E) Palmar and dorsal interossei.

(F) Lumbricals.

(B) Hypothenar muscles -

1. Abductor digiti minimi,
2. Flexor digiti minimi, and
3. Opponens digiti minimi.

Nerve supply of these muscles :

I. Median nerve supplies,

1. Thenar muscles.
2. Two lateral lumbricals.
3. Sometimes, 1st dorsal interosseous muscle.

II. Ulnar nerve supplies,

1. Hypothenar muscles.
2. All palmar and dorsal interossei.
3. Two medial lumbricals.
4. Two heads of adductor pollicis.
5. Medial head of flexor pollicis brevis (thenar muscle).
6. Palmaris brevis.

N.B. : Ulnar nerve supplies all the small muscles of hands excepting the thenar muscles and the two lateral lumbricals.

How to diagnose wasting of the small muscles of hands ?

1. Flattening of palm due to wasting of thenar and hypothenar muscles.
2. Prominent knuckles and bony prominences.
3. Flexor-extensor tendons will stand out.
4. The interosseous space in the dorsum of hand will be hollowed or depressed due to wasting of interossei and lumbricals.

Test of interossei and lumbricals :

Test the patient's ability to flex all of his MCP joints and to extend all the DIP joints of hand. Remember, the palmar interossei are also adductors and dorsal interossei are abductors of fingers.

Test of 1st dorsal interosseous — Place the flat of the hand with separated fingers over a table and ask the patient to abduct his index finger against your resistance. The muscle can be seen and felt to contract.

How to test these small muscles ?

I. Abductor pollicis brevis— The patient is asked to abduct his thumb in a plane at right angles to the palmar aspect of index finger, against resistance offered by your thumb. The muscle can be felt to contract. This muscle is affected first in carpal tunnel syndrome.

II. Adductor pollicis— Give the patient a book ('book test') and ask him to grasp it firmly between the thumb and other fingers of both hands. In a normal subject the thumbs remain straight whereas in lesions of the ulnar nerve, the terminal phalanx of the thumb of the affected hand will be flexed (Froment's sign).

III. Opponens pollicis— Ask the patient to touch the tip of all the fingers with his thumb. You can oppose the movement with your thumb or index finger, or you can ask the patient to swing the thumb across the palm.

Common causes of wasting of the small muscles of hand :

THE SMALL MUSCLES OF THE HAND ARE SUPPLIED BY C₈ AND T₁ SEGMENTS OF THE SPINAL CORD. The causes of wasting, therefore, include lesions in the lower motor neurones at any point between these spinal segments and the muscles, together with certain other conditions in which primary muscle degeneration or reflex muscular wasting occurs.

a) Lesion in the anterior horn cells—

1. Acute anterior poliomyelitis.
2. Motor neurone disease (amyotrophic lateral sclerosis).
3. Syringomyelia.
4. Intramedullary tumours like glioma, ependymoma.

b) Lesion in the nerve roots (anterior roots) or radiculopathy—

1. Extramedullary lesion, e.g., patchy arachnoiditis.
2. Cervical spondylosis.
3. Neuralgic amyotrophy.
4. Leptomeningitis (syphilitic) - Rare.

c) Lesion in the spinal nerve — Klumpke's paralysis from birth injury.**d) Lesion in the brachial plexus —**

- | | |
|-----------------------------|----------------|
| 1. Cervical rib. | 3. Trauma. |
| 2. Thoracic inlet syndrome. | 4. Metastasis. |

e) Lesion in the median and ulnar nerve —

1. Injury or trauma.
2. Peripheral neuropathy from leprosy, lead neuritis etc.
3. Peroneal muscular atrophy (usually follows wasting of legs and feet).
4. Carpal tunnel syndrome.

f) Muscle disease and others —

- | | |
|----------------------------------|--------------------------------------|
| 1. Distal myopathy of Gower's. | 3. Volkmann's ischaemic contracture. |
| 2. Myotonia dystrophica, rarely. | 4. Peripheral vascular disease. |

g) Reflex wasting (due to disuse atrophy) — Rheumatoid arthritis, post-paralytic, post-fracture.**h) Systemic wasting —** Malignancy, tuberculosis, diabetes mellitus, thyrotoxicosis, AIDS.

N.B. : Five common causes of wasting of the small muscles of hands are,

1. Leprosy.
2. Motor neurone disease.
3. Rheumatoid arthritis.
4. Poliomyelitis.
5. Syringomyelia.

Wasting of small muscles of both upper and lower extremities :

1. Charcot-Marie-Tooth disease and other motor peripheral neuropathies.
2. Distal myopathy of Gower's.
3. Distal spinal muscular atrophy.
4. Post-G.B. syndrome.
5. Myotonia dystrophica.
6. Rheumatoid arthritis.

How to diagnose cervical rib clinically ?

The patient may complain of pain along the ulnar border of forearm and hand. Horner's syndrome may be evident. Adson's test confirms the presence of cervical rib.

Adson's test — This test is also positive in thoracic inlet syndrome. The examiner feels the radial pulse from the back of the sitting patient. Now the patient is asked to take a deep breath and to turn the face (not flexion) towards the affected side. In the presence of cervical rib, there will be obliteration or diminution of pulse.

Identification points for an aetiological diagnosis :

1. History from childhood -
 - a) Poliomyelitis.
 - b) Klumpke's paralysis.
2. If complains of neck pain -
 - a) Cervical spondylosis.
 - b) Intra- or extramedullary tumour.
 - c) Cervical rib (may be associated with bony lump).

3. If associated with fasciculation and brisk tendon reflexes in upper extremities — Amyotrophic lateral sclerosis.
 4. Associated with painful swelling of joints - Rheumatoid arthritis. ■
 5. If there are **trophic changes** in hand-
 - a) Leprosy.
 - b) Syringomyelia.
 - c) Cervical rib.
 6. Examine the elbow for signs of old injuries like excessive callus, cubitus valgus deformity (for nerve injury) or for **thickened ulnar nerve** (leprosy).
 7. If there is :
 - a) Absence of sensory loss - MND.
 - b) Dissociation of sensory loss - Syringomyelia.
- N.B. : Wasting of the small muscles of hands may be associated with claw hand.

Case 53

Claw Hand

Describe the claw hand deformity ;

Claw hand or 'main-en-griffe' is a condition where the metacarpophalangeal (MCP) joints are hyper-extended. and the PIP and DIP joints are flexed.

We know that the lumbricals are the main flexors of the first phalanx and the interossei are the sole extensors of the middle and distal phalanges. When the lumbricals are paralysed, there is hyperextension at MCP joints (by unopposed action of extensor digitorum) and when the interossei are paralysed, the PIP and DIP joints are flexed. So **claw hand is produced by the paralysis of interossei and lumbricals.**

Enumerate the common causes of claw hand :

The interossei and lumbricals are supplied by the T₁ segment of the spinal cord through the ulnar and median nerves. So any lesion of T₁ segment or of the nerves will produce the claw hand deformity.

1. True claw hand :

- a) Combined lesion of ulnar and median nerves by injury or leprosy (tuberculoid variety) will produce claw hand. It is obvious that only ulnar nerve affection will produce ulnar claw hand.
- b) Cervical rib (by friction against the lowest trunk of the brachial plexus) or thoracic inlet syndrome.
- c) Klumpke's paralysis.
- d) Motor neurone disease (progressive muscular atrophy, amyotrophic lateral sclerosis), syringomyelia, intramedullary tumours etc.

N.B. : Actually the diseases which produce wasting of the small muscles of hands may show some degree of claw deformity.

2. Mimicking (pseudo) claw hand :

These are basically surgical conditions like :

- a) Dupuytren's contracture.
- b) Volkmann's ischaemic contracture.
- c) Post-burn contracture.

Aetiological differentiation by sensory functions :

1. Motor neurone disease, either amyotrophic lateral sclerosis or progressive muscular atrophy — No sensory loss.
2. Injury to the ulnar and median nerve -
 - a) Practically, all modalities of sensation in the palm and the dorsum of hand are lost.
 - b) In the dorsum of hand, a small area escapes sensory loss (over the base of thumb and the first interosseous space) as it is supplied by radial nerve.
3. Leprosy — Patchy and distal sensory involvement (peripheral neuropathy).
4. Syringomyelia — Dissociated sensory loss, i.e., loss of pain and temperature sensation with preservation of fine touch sensation.

Hand feels very cold : what will you think ?

It is the vasomotor insufficiency produced by the presence of cervical rib. thoracic inlet syndrome or due to severe form of vasculitis. The affected hand feels colder than the normal side, and becomes pale when elevated and blue when dependent for some time.

What is wrist drop ?

(A) Definition . Paralysis of the extensors of wrist. There will be difficulties in extension of wrist and fingers (finger drop). In an attempted extension of fingers, there will be flexion of MCP joints and extension of interphalangeal joints due to unopposed action of lumbricals and interossei.

(B) Method of testing : Ask the patient to make a fist and try to flex the wrist forcibly against patient's effort to maintain the posture. Normally, it is almost impossible to flex the wrist overcoming the patient's wrist extensors. But in wrist drop, patient's wrist will be very easily flexed by the examiner.

(C) Aetiology :

1. Radial nerve palsy (trauma, Saturday night palsy or compression at axilla),
2. Chronic lead poisoning, and
3. Other peripheral neuropathies (including mononeuritis multiplex).

Identification points for an aetiological diagnosis :

Same as described in the section on 'Wasting of the small muscles of hands'.

*** Never forget to palpate the ulnar nerve in the elbow (e.g., leprosy) and to examine for trophic changes in the fingers in a patient with claw hand or wasting of the small muscles of hands.**

Case 54**GYNAECOMASTIA (GYNAECOMAZIA)****What is gynaecomastia ?**

This is a condition where the male breast looks like that of a female. On inspection, even the nipple and areola assume a feminine appearance. Early gynaecomastia is characterised by proliferation of both the fibroblastic stroma and the duct system. Eventually the number of ducts decrease. It is usually painless but elderly person with gynaecomastia may have little pain. Gynaecomastia results from an increase in the oestrogen/androgen ratio.

What is lipomastia ?

Lipomastia or 'false' enlargement of breast (found in obesity) is best palpated with flat of the palm while gynaecomastia is best appreciated with fingers. In lipomastia, no breast tissue is there but only mammary fat is present. One should remember that no breast tissue is palpable in normal man.

* Pseudogynaecomastia is found in lipomastia, neurofibromatosis, cold abscess or malignancy.

** Painful gynaecomastia : Puberty or adolescence, drug-induced (e.g., spironolactone) and cirrhosis of liver.

Examination of the patient :

1. Examine the breast tissue first with flat of the palm, then with fingers to confirm the glandular tissue. Tenderness indicates rapid growth.
2. Age of the patient.
3. Unilateral or bilateral.
4. **Stature** of the patient and signs of eunuchoidism.
5. H/O taking drugs (produce gynaecomastia) or alcoholism.
6. H/O mumps or castration.
7. Examine both the **testes**.
8. Examine the **liver, spleen**, and search for any hepato-celular failure.
9. Look for the signs of carcinoma of the lung in the form of SVC obstruction, collapse of the lung.
10. Examine the spine for any tenderness or gibbus (caries spine).
11. Signs of leprosy nodules, hypopigmented spots in skin or thickened peripheral nerves etc.
12. P/R examination to exclude carcinoma of the prostate for which stilboestrol is advised.

Drugs *producing gynaecomastia :*

1. Oestrogens.
2. Digitalis (due to oestrogen-like effect).
3. Spironolactone (reduces serum testosterone level)— Often the breasts are tender.
4. Cimetidine (anti-androgenic).
5. INH (cause not known).
6. Methyldopa (cause not known).
7. Marijuana, cannabis or heroin abuse (associated with high oestrogen and low testosterone).
8. Busulphan, phenothiazines, tricyclic antidepressants, ketoconazole, diazepam, phenytoin.

Explanation of your method of examination :

1. Age of the patient— Newborn, adolescence and old age may have physiological gynaecomastia.
2. Stature of the patient— Klinefelter's syndrome and hypogonadism may have tall stature.
3. Drugs, mumps orchitis, castration— all produce gynaecomastia.
4. Examination of testes :
 - " a) Testicular tumour— Asymmetry of testes, smooth or lobulated (e.g., Leydig cell tumour).
 - b) Klinefelter's syndrome— Small and firm testes.
 - c) Cirrhosis of liver with hepato-cellular failure— Small and soft testes.
 - d) Lepromatous leprosy— Atrophied small and soft testes.

* By Prader orchidometer, testicular size < 3 cm indicates atrophy (normal adult size is 3.5-5.5 cm).

Pre-pubertal males have small and rubbery testes.

5. Examination for hepatosplenomegaly :
 - a) Cirrhosis of liver.
 - b) Leprosy.
 - c) Acute leukaemia (leukaemic deposits in breast may mimic gynaecomastia).
6. Facies :
 - a) Puffy, bloated and congested— SVC obstruction due to carcinoma of the lung.
 - b) Leonine facies— Lepromatous leprosy.
 - c) Hepatic facies— Cirrhosis of liver with hepato-cellular failure.
 - d) Rarely true hermaphroditism.
7. Spine— Cold abscess may form behind the breast tissue in a patient suffering from caries spine (abscess coming in front through muscle plane). It is not true gynaecomastia and usually produces unilateral painless swelling of the breast.
8. Examination of the skin is done for hypopigmented anaesthetic spots, fall of body hairs and eyebrows, and subcutaneous nodules for lepromatous leprosy which causes gynaecomastia. Sparse pubic hair is common in hypogonadism.
9. P/R examination — If the patient gives history of taking stilboestrol (probably suffering from carcinoma of the prostate).
10. Unilateral or bilateral— Gynaecomastia may be unilateral to start with. Klinefelter s syndrome and physiological gynaecomastia are bilateral.

Common causes of gynaecomastia :

1. Physiological— Newborn, adolescence and old age.
2. Klinefelter's syndrome (47, XXY).
3. Hepato-cellular failure commonly from alcoholic cirrhosis.
4. Drug-induced.
5. Bronchogenic carcinoma.
6. Lepromatous leprosy.
7. Leukaemic deposits in breasts (not true gynaecomastia).
8. Starvation; recovery from wasting diseases.
9. Hypogonadism due to any cause (secondary testicular failure from orchitis or castration).
10. Obesity.
11. Oestrogen-secreting testicular (Sertoli cell tumour) or adrenal tumours.
12. Hyperthyroidism, renal diseases, hyperprolactinaemia, local trauma.

How to proceed for diagnostic evaluation ?

After proper clinical evaluation :

1. A careful drug history is taken.
2. Measurement and examination of testis (if both are small and firm, a karyotype is indicated; if testes are asymmetric, an evaluation for testicular tumour is warranted).
3. Liver function tests.
4. Blood for testosterone, LH, FSH, oestradiol, prolactin and HCG estimation.
5. 24-hours urinary 17-ketosteroid estimation.

Explanation : If there is,

- LH T and testosterone 4- = testicular failure.
- LH J. and testosterone i = most likely there is increased primary oestrogen production (e.g. Sertoli cell tumour of the testis).
- LH T and testosterone T = androgen resistance or gonadotropin-secreting tumour.
- T Urinary 17-ketosteroid = feminising adrenal tumour.

Work up is necessary where,

- a) Drug history is negative.
- b) Breast is tender with rapid growth.
- c) Breast mass > 4 cm in diameter.

How do you like to treat your patient :

1. Reassurance for pubertal gynaecomastia (may persist for 2-3 years).
2. Treatment of the cause like leprosy, hepato-cellular failure, withdrawing the offending drug etc.
3. Painful gynaecomastia who are not candidates for other therapy — Treatment with antioestrogens, e.g., tamoxifen (20 mg/day) should be tried. Aromatase inhibitors (testolactone can be effective). Patients with testicular insufficiency may be benefited by androgen replacement therapy.
4. Indication for surgery (simple mastectomy or liposuction) —
 - (i) Severe psychological problem.
 - (ii) Cosmetic problem.
 - (iii) Continued growth.
 - (iv) Suspicion of malignancy.

* In gynaecomastia, the risk of developing carcinoma of the breast is very very rare.

Case 55**Hypo- or Hyperpigmentation****Common causes of hyperpigmentation**

- | | |
|----------------------------|--|
| 1. Familial, racial. | 10. Chronic arsenical poisoning. |
| 2. Sunburn. | 11. Facial pigmentation in chloasma, SLE. |
| 3. Addison's disease. | 12. Porphyria (cutanea tarda). |
| 4. Haemochromatosis. | 13. Freckles, lentigos (post-sunexposure). |
| 5. Peutz-Jeghers syndrome. | 14. Urticaria pigmentosa. |
| 6. Chronic kala-azar. | 15. Acanthosis nigricans. |
| 7. Cirrhosis of liver. | 16. Melanoma. |
| * 8. Drug-induced. | 17. Scleroderma. |
| 9. Pellagra. | 18. Paraneoplastic syndrome. |

* Clofazimine (reddish-brown), busulphan, oral contraceptive pills, bleomycin, phenothiazines, heavy metals (arsenic, gold), tetracycline, amiodarone, minocycline, psoralens.

** Abnormal increased pigmentation is known as hyperpigmentation which may be localised (commonly due to dermatological disorders) or generalised (commonly due to systemic diseases).

Causes of hypopigmentation :

Read the section on 'Pityriasis versicolor'.

Sites examined for increased pigmentation :

1. Face.

2. Inside the oral cavity, specially buccal mucosa (commonly seen in Addison's disease, malabsorption syndrome, chronic cachexia, Peutz-Jeghers syndrome and haemochromatosis), palate and tongue.
3. Palmar creases.
4. General skin surface (pressure points; normally pigmented areas like areola, genititalia, sun-exposed areas; recent scars); palms and soles.

Pigmentation in chronic kala-azar :

Due to increased black pigmentation, the disease is known as kala-azar (the Hindi version of 'black' is 'kala'). Hyperpigmentation occurs as a result of disturbance in melanin metabolism. Pigmentation is commonly seen **around the mouth, on the temple and forehead.**

Pigmentation in Addison's disease :

Pigmentation in Addison's disease is due to increased pituitary MSH and ACTH secretions. At first, there is tanning noticed following sun exposure. Afterwards, there is increased pigmentation **at pressure points** (elbow), in the **areas normally pigmented** (areola, genitals, knuckles, palmar creases, old and new scars) and also in the **mucous membranes** (mouth and genitals). Rarely, there may be areas of vitiligo.

Features of addison's disease :

Commonly the patient is female and suffers from :

- | | |
|--------------------------------------|-------------------------------------|
| 1. Asthenia (glucocorticoid 4) | I
V
J cardinal manifestations |
| 2. Hypotension (mineralocorticoid 4) | |
| 3. Hyperpigmentation (ACTH t) | |
| 4. Anorexia, malaise, weight loss. | |
| 5. Decreased body hairs. | 8. Nausea, vomiting. |
| 6. Hyperkalaemia. | 9. Chronic fatigue syndrome |
| 7. Hypoglycaemia. | 10. Acute adrenal crisis. |

* Thomas Addison (1793-1860) was a physician in Guy's Hospital, London, UK.

Differentiation between primary and secondary adrenocortical hypofunction :

Glucocorticoids, mineralocorticoids and androgens are low in both the conditions.

In primary (Addison's disease) cases, there are features of,

1. Pigmentation,
2. Absence of hypothyroidism and hypogonadism, and
3. High plasma ACTH level.

In secondary hypofunction (in involvement of pituitary or hypothalamus), there are :

1. Absence of pigmentation.
2. Presence of hypothyroidism and hypogonadism, and
3. Low plasma ACTH level.

What is haemochromatosis ?

It is an iron-storage disorder with inappropriate increase in intestinal iron absorption. There is deposition of iron in parenchymal cells with eventual tissue damage. The disease is clinically manifested by,

1. Excessive generalised skin pigmentation (due to increased melanin and iron deposition; melanin gives the typical bronzing or slaty grey hue of the skin).
2. Diabetes mellitus (bronze-diabetes).
3. Arthropathy.
4. Cardiac involvement — CCF, cardiomyopathy.
5. Hepatic involvement (cirrhosis or hepato-cellular carcinoma) with hepato-cellular failure.
6. Loss of libido and testicular atrophy.

* The **pigmentation is more pronounced** on face, neck, lower forearms, dorsa of hands, lower legs, genitals and in scars.

Treatment of haemochromatosis :

1. Phlebotomy. "
2. Iron chelation by desferrioxamine.
3. Treatment of CCF, hepato-cellular failure, diabetes mellitus etc.

How a haemochromatosis patient dies ?

1. CCF.
2. Hepato-cellular failure.
3. Hepato-cellular carcinoma (in treated cases).

What is Peutz-Jeghers syndrome ?

In this syndrome, there are numerous polyps seen in small and large intestine. The polyps show features of hamartoma on histological section. Chance of malignant transformation is very rare. The syndrome is associated with mucocutaneous pigmentation and rarely, germinal cell tumour of ovary. Commonly it gives rise to **perioral pigmentation**.

What is chloasma or melasma ?

A common hypermelanosis of young females (very rare in males) frequently precipitated by pregnancy (chloasma gravidarum) or by prolonged use of oral contraceptive pills (also known as 'mask of pregnancy'). The pigmentation is generally seen on malar prominences and bridge of the nose (bilateral and frequently symmetrical); also seen in forehead and moustache area. It is a benign condition.

What is acanthosis nigricans ?

It is a thickened, velvety hyperpigmentation predominantly of the flexures (e.g., sides of neck, axilla). In majority, it is associated with obesity who is also having underlying insulin resistance. It can be a reflection of internal malignancy (e.g., G. I. tract), or endocrinopathy such as acromegaly, Cushing's syndrome or insulin-resistant diabetes mellitus.

Causes of 'plethoric face' ?

- | | |
|------------------------|---------------------------|
| 1. Chronic alcoholism. | 4. SVC syndrome. |
| 2. Cushing's syndrome. | 5. Chronic cor pulmonale. |
| 3. Polycythemia. | 6. Carcinoid syndrome. |

* See the section on 'Mitral stenosis' for causes of 'malar flush'.

Case 56

SCABIES

Aetiology and transmission :

This contagious disease is caused by the mite *Sarcoptes scabiei* contracted through close personal contact or contaminated clothings. After copulation the fertilised female mite excavates a burrow in the stratum corneum. From egg — nymphs hatch out — mature into adult males and females on the surface of the skin — mate, and reinvade the skin of same or other person.

Doctors, paramedicals and inmates of mental hospitals are at particular risk to have scabies.

Characteristic distribution :

1. Webs of fingers, wrist, ULNAR BORDER OF FOREARM and ARM, anterior axillary folds.
2. Front of chest, areola in female breast, UMBILICAL and PERIUMBILICAL REGION, belt line and lower abdomen.
3. SHAFT OF PENIS, SCROTUM, vulva, folds of buttock, GROIN, MEDIAL ASPECT OF THIGH, knee, front of ankle, webs of toes.

Spread is by prolonged close contact, sharing of clothings and beddings, and by sexual exposure.

An imaginary circle intersecting the main sites of involvement—axilla, elbow flexures, wrist, hand and groins has long been called the 'circle of Hebra'.

Which part of body is usually spared ?

Face and scalp (face and scalp may be affected in infants).

How to diagnose scabies ?

1. Specific lesion with typical distribution.
2. Burrow.

3. Nocturnal pruritus (intractable).
4. H/O affection of several members of the family at a time (as scabies is highly contagious).
5. Demonstration of mite *Sarcoptes scabiei* from the burrow (confirmation of diagnosis).

* Itching from scabies also increases after a hot bath.

Most pathognomonic lesion of scabies :

Burrow or Tunnel (linear or curved) is the most pathognomonic and characteristic lesion.

This is an irregular or linear skin-coloured ridge caused by tunneling of the mite in the stratum corneum layer of skin. It is seen in areas mentioned under distribution of lesion. The burrow can be identified by 'ink method' (touched with a drop of ink) when the burrow absorbs the ink and is highlighted as a dark line.

Commonest lesion in scabies :

Papulo-vesicles.

Describe the different lesions :

1. Papules (itchy and red).
2. Vesicles,
3. Pustules,
4. Impetigo,
5. Burrow, and
6. Indurated nodules.

Mode of spread :

1. Very close and intimate contact with scabies patient (e.g., sharing the bed)
2. Overcrowding.
3. Poverty.
4. Poor hygiene.
5. Sexual promiscuity.

Why there is nocturnal itch ?

When the patient retires in warm bed, the mites start moving within the burrow.

Non-itchy scabies :

1. Norwegian scabies, or
2. Scabies with psychiatric illness.

What is Norwegian (crusted) scabies ?

Widespread hyperkeratotic and crusted lesions of scabies (extremely infectious) resulting from infestation with thousands or millions of mites, and are present in :

1. Mentally retarded patients (e.g., schizophrenia, Down's syndrome),
2. Patients with poor cell-mediated immunity (e.g., lepromatous leprosy, AIDS),
3. Severely debilitated patient (e.g., severe malnutrition), and
4. Immunocompromised states e.g., diabetes mellitus, glucocorticoid therapy, organ transplantation.

Lesions present in face :

1. Neonates, or
2. Norwegian scabies.

D/D of scabies :

- | | |
|--------------------------|------------------------------|
| 1. Pediculosis corporis. | 3. Papular urticaria. |
| 2. Atopic dermatitis. | 4. Dermatitis herpetiformis. |

Mention some itchy lesions in skin :

- | | |
|------------------------------|--------------------------------------|
| 1. Scabies. | 7. Eczematous and atopic dermatitis. |
| 2. Dermatitis herpetiformis. | 8. Pediculosis. |
| 3. Urticaria. | 9. Ringworm. |
| 4. Lichen planus. | 10. Seborrhoeic dermatitis. |
| 5. Insect bite. | 11. Drug eruption. |
| 6. Psoriasis. | 12. Xerosis (dry skin). |

Medical causes of generalised pruritus :

1. Obstructive jaundice (medical or surgical).
2. Diabetes mellitus.
3. Chronic renal failure.
4. Polycythemia vera (specially after a hot bath).

5. Systemic mastocytosis.
6. Lymphoma (Hodgkin's disease commonly) and leukaemias; multiple myeloma.
7. Carcinoid syndrome.
8. Thyrotoxicosis.
9. Drugs like cocaine, morphine.
10. Psychogenic.
11. Drug reaction, pregnancy (last trimester).
12. HIV infection.
13. Neurosis.
14. Old age, specially in winter; dry skin.
15. Malignancy.

Complications of scabies :

1. Pyoderma, specially impetigo and ecthyma.
2. Acute glomerulonephritis— Due to sensitisation by nephritogenic strains of streptococci (most dangerous).
3. Eczema— Sensitization from parasites or from prolonged itching.
4. Rarely, sulphur dermatitis after initiation of treatment.
5. Secondary lymphadenitis.

* Always search for signs of scabies infection in a patient with acute glomerulonephritis.

Management :

1. At first, the treatment of secondary infection is done by antibiotics. Patient should cut his nails short and his daily used articles (towel, pillow, bed-sheet) should be kept separate and laundered properly. Antihistaminics and calamine lotion may be necessary to control itching. Asymptomatic family members should be treated simultaneously.
2. Instructions to the patient— Start with a bath and dry thoroughly afterwards. Apply the medicine thinly but thoroughly over the whole body from chin downwards (both involved and uninvolved areas). It is better to rub the medicine into the skin. Treatment is best done at night before going to bed. Avoid touching mouth or eyes with hands. Change the undergarments and sheets on the next day and launder them. The patient may take a bath in the morning with soap and water.

A single overnight application with permethrin is sufficient. In pregnant women and infants, sulphur ointment, permethrin and crotamiton are preferred. Benzyl benzoate needs 3 applications at 12-hourly interval without intervening bath. Sulphur ointment is used once daily for consecutive 3 days. Do not repeat the drugs unless instructed.

3. Medicines used :

- | | | |
|---|---|--------------------------------|
| <ol style="list-style-type: none"> a) Benzyl benzoate lotion (25%) or emulsion (child 12.5%) b) Unguentum sulphur (adult 10%, child 5%) c) Gamma benzene hexachloride (1%) as lotion or cream. d) Crotamiton (10%) as lotion or cream. e) Mitigal - 10% solution in liquid paraffin. f) Monosulfiram (25%; tetmosol)— used as soap or lotion. g) Topical thiabendazole. h) Permethrin (5%) — used as cream; single application, may be repeated after 1 week. i) Malathion (0.5%) lotion. j) Ivermectin (200 (ig/kg, orally in single dose; adult dose in 12 mg) effectively treat scabies. | } | apply for consecutive 2 nights |
|---|---|--------------------------------|
- Two or more doses at 1 week apart are required for crusted scabies.

* *Itching in scabies* is due to development of sensitization to some of the products (e.g., saliva-scabin) of the mite.

How to diagnose (confirm) scabies under the microscope :

A selected lesion is scraped by sterile needle or scalpel 6-7 times and the scrapings are transferred to a glass slide on which 10% KOH solution has already been kept. The slide is then examined with low power magnification for mites, eggs or faecal pellets.

Case 57

PSORIASIS

Describe the lesion :

1. Red, scaly papules or plaques.
2. Sharply defined.
3. Covered with varying amount of loosely adherent silvery-white scales.
4. Present over knees, elbows, hands, lumbosacral region, scalp, rarely in palms and soles (i.e., MAINLY OVER THE EXTENSOR SURFACES).
5. The lesions are pruritic.
6. Localised lesion to sites of trauma (**Koebner's phenomenon**).
7. Scraping the scales leaves behind punctate bleeding spots (**Auspitz's sign**) - DIAGNOSTIC.

* Usually in young adults, both sexes with genetic predisposition; unpredictable course with relapses and remissions; worse in winter season; no available test to predict outcome.

What is psoriatic arthropathy ?

Arthritis occurs in 5-10% patients of psoriasis. The five distinct clinical forms of arthritis are :

1. Asymmetrical oligoarthritis (35%).
2. Symmetrical seronegative arthritis (30%)—'rheumatoid arthritis pattern'.
3. Arthritis of **distal interphalangeal joint** (15%)—most characteristic.
4. Sacroiliitis and/or spondylitis (15%).
5. Arthritis mutilans — Severe destructive arthritis with deformities of hands and feet (5%).

There may be associated nail changes; rheumatoid factor is negative. Arthritis is commonly associated with HLA-B27 allele. Arthropathy may progress to deformity of fingers (arthritis mutilans may lead to 'telescoped' phalanges).

Clinical forms of psoriasis :

1. Plaque psoriasis— Commonest lesion; coin shaped (numular);
2. Guttate—Raindrop-like, small-sized.
3. Palmo-plantar— Often with sterile pus in palms and soles.
4. Erythrodermic form— Exfoliative dermatitis; may be fatal.
5. Genital— Penis and vulva affected.
6. Flexural (intertriginous)— Instead of extensor surfaces, the flexural sites are involved.
7. Rupoid— Hyperkeratotic, cone-shaped.
8. Scalp psoriasis— Fairly common; not associated with alopecia.
9. Generalised pustular— Known as 'Von Zumbusch' psoriasis; rare but very serious.

* *Exfoliative dermatitis (erythroderma)* is common in psoriasis, contact dermatitis, atopic dermatitis, pityriasis rubra pilaris, drug-induced (e.g., sulphonamides, dapsone) and systemic diseases (lymphoma/ carcinoma/multiple myeloma/HIV infection).

Describe the nail changes in psoriasis :

Nail changes are often associated with arthropathy and may reflect the severity of the disease.

1. Pitting (thimble pitting),
2. Subungual hyperkeratosis,
3. Destruction of nail plates,
4. Discolouration (yellow-brown) of nail plates, and
5. Onycholysis (separation of nail from nail-bed) and horizontal ridging.

* Onycholysis is the most characteristic nail lesion in psoriasis.

Causes of pitting nail :

1. Psoriasis.
2. **Eczema.**
3. Alopecia areata.

Diseases with positive Koebner's phenomenon :

1. Psoriasis.
2. Lichen planus.
3. Viral **wart**.

Describe macule and papule ?

(A) Macule - Localised area of change of colour in skin, neither raised nor depressed (non-palpable lesion). A big macule is known as 'patch'.

(B) Papule - Raised from skin surface and generally < 5 mm in size; big papule is known as 'plaque'.

* Vesicle is small (< 5 mm), usually clear fluid-containing elevated lesion whereas bulla are large vesicle (> 5 mm). Nodules are solid elevation in skin of > 5 mm in diameter.

Histology of skin lesion in psoriasis :

- | | |
|---|--|
| 1. Parakeratosis (immature cells in stratum comeum) | } reflecting increase in skin turnover |
| 2. Acanthosis (hyperplasia of stratum malpighii). | |
| 3. Loss of granular cell layer. | |
| 4. Dilated and tortuous blood vessels in dermal papillae. | |
| 5. Rarely, neutrophilic abscesses (microabscesses of Munro) in the horny layer. | |

Investigations you like to perform :

1. Blood for TC, DC to exclude superadded infections.
2. Skin scrapings and nail clippings may be examined for tinea (for differential diagnosis from fungal infection).
3. Rheumatoid factor - Negative (for differential diagnosis of arthropathy).
4. X-ray of affected joints may show 'pencil-in-cup' deformity.
5. Skin biopsy may be done (seldom necessary because the clinical picture is very characteristic).

Management :

The treatment is concerned with control rather than cure and is divided into four categories :

- Topical agents (emollients)—'weak' tar, corticosteroids, vitamin D., agonists or dithranol preparations.

• PUVA or PUVB therapy.

- Systemic agent—retinoids (tazarotene), immunosuppressive drugs (cyclosporin A) or newer biological agents (infliximab).

e Intensive treatment with topical agents and UV-rays under medical supervision (No. 6 below).

1. Coal-tar preparations with keratolytics (salicylic acid 3-5%) as an ointment.
2. Dithranol (0.1%) cream—used locally; it inhibits DNA synthesis.
3. Corticosteroid ointment may be used locally (systemic corticosteroid is strictly contraindicated).
4. PUVA therapy (photochemotherapy) — 'P' stands for Psoralen taken orally and UVA stands for ultra violet light-A, i.e., of long-wavelength (320 - 400 nm). Oral psoralen is followed 2 hours later by UVA therapy; 2-4 sittings / week are necessary (total 10-20 sittings). UVB is helpful in winter season.
5. Topical preparations of retinoic acid derivatives (0.1%) may be tried in localised plaques.
6. Topical therapy with ultraviolet rays :
 - a) Goeckerman regimen—tar plus UVB.
 - b) Ingram regimen—dithranol plus UVB.
7. Methotrexate—Used in severe erythrodermic form, arthropathy or resistant cases. Methotrexate is used once weekly in a dose of 5 mg BDPC (used over 24 hours), and is supplemented by folic acid therapy (5 mg, once weekly except the day of methotrexate).
8. Calcipotriol (vitamin D₃ analogue)—Applied twice daily, not exceeding 100 g/week.
9. NSAID for arthropathy.
10. Nail dystrophy is resistant to treatment.

* Oral corticosteroid may be used judiciously in refractory psoriasis, psoriatic arthropathy and erythrodermic form. Flexural psoriasis is treated with calcitriol (vitamin D₃ analogue) and 0.1% tacrolimus ointment. Resistant cases may be treated with cyclosporin A, acitretin or hydroxyurea. TNF-α blockers or biologics (infliximab, etanercept, efalizumab) are now being tried with success.

** UVC : Wavelength 10-290 nm (does not reach earth), UVB : 290-320 nm. UVA : 320-400 nm, visible white light we see : 400-700 nm, infrared rays (primarily evoke heat) > 700 nm.

Conclusion :

Always examine the nails and joints in a patient of psoriasis. Psoriasis does not affect the mucous membrane. One should not forget to examine the elbows, knees, back of the trunk and scalp.



Arachnodactyly – long and slender fingers, compared with a normal-sized hand



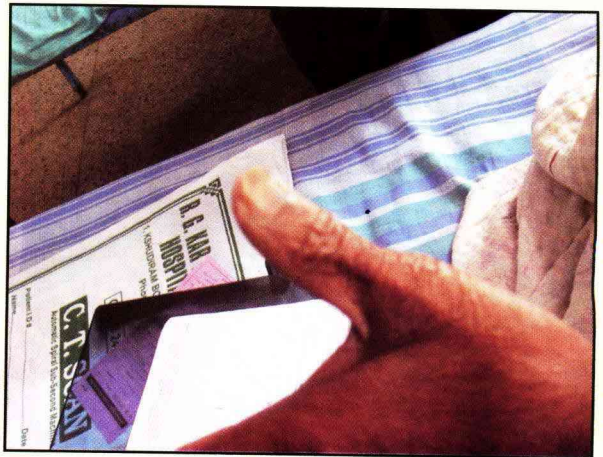
Rheumatoid nodule on extensor surface of upper part of forearm



Digital clubbing seen in cyanotic congenital heart disease



Rheumatoid arthritis hand characterized by synovial swelling at the wrist, swelling of MCP and PIP joints, Z-deformity of thumb, and swan neck deformity of fingers with mild ulnar deviation of hand



Koilonychia in a middle-aged patient with bleeding piles (resulting in chronic iron deficiency anaemia)



A case of **meningitis** on the day of admission with discharging right ear



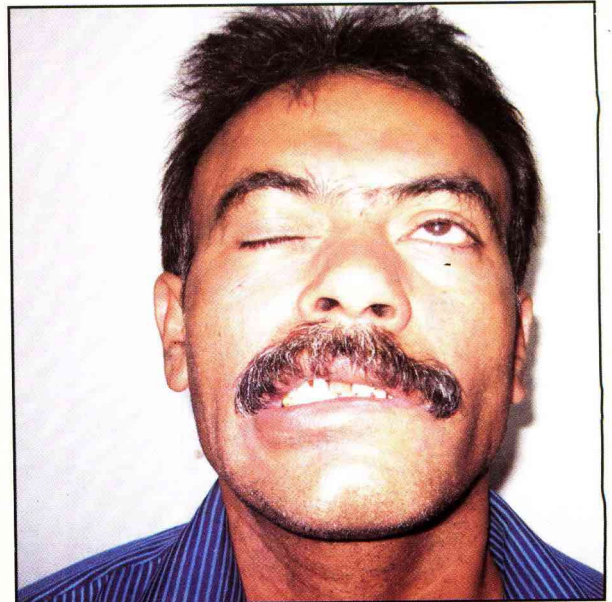
Bilateral partial ptosis (right > left) in **myasthenia gravis**



Squint developed as a result of left-sided **VIth nerve palsy**. Here, it is a false localizing sign due to raised intracranial tension



Gowers' sign in Duchenne type of muscular dystrophy



Left-sided **Bell's palsy**. Deviation of face to the right side occurs on showing the teeth as well as Bell's phenomenon is noted on forced closure of eyes

Case 58

PITYRIASIS VERSICOLOR (TINEA VERSICOLOR)

Characteristics :

1. Caused by *Malassezia furfur* (*M. furfur* is possibly the parasitic form of a common saprophytic yeast, often called *Pityrosporum orbiculare*).
2. Occurs in warm and humid climates; a relatively common condition affecting young adults.
3. Well-defined, hypopigmented small macules with fine branny scales (the lesion is of various size, shape and colour).
4. Generally non-itchy.
5. Many small spots may coalesce and form a large depigmented spot.
6. Classical sites are front of the chest, back of the chest, sides of neck, upper arm and face.
7. Sweating aggravates the condition; diabetes mellitus and pregnancy may be predisposing factors.
8. Asymptomatic except the cosmetic problem.

* The fungal hyphae and budding yeast may be seen in the scraped scales when treated by 10% KOH solution and seen by light microscopy: on inspection under Wood's lamp, it gives yellow fluorescence.

D/D of hypopigmented (hypomelanotic) spots :

1. Tuberculoid leprosy (test for touch sensation and examine for thickened nerve).
2. Vitiligo (may be idiopathic, sometimes associated with autoimmune diseases).
3. Pityriasis alba (found in the face in children; probably infective; asymptomatic, remits spontaneously).
4. Post-burn (past H/O burn) or post-inflammatory.
5. Tuberous sclerosis (H/O convulsions, low intelligence and adenoma sebaceum in face).
6. Prolonged chloroquine therapy (H/O prolonged chloroquine intake, e.g., rheumatoid arthritis, SLE).

* Others ; pityriasis versicolor, psoriasis, albinism, nevus depigmentosus, piebaldism and idiopathic guttate hypomelanosis.

Common scaly lesions in skin :

- | | |
|----------------------------|---|
| 1. Psoriasis. | 5. Certain eczema. |
| 2. Pityriasis rosea. | 6. Exfoliative dermatitis (erythroderma). |
| 3. Seborrhoeic dermatitis. | 7. Ringworm. |
| 4. Lichen planus. | 8. Ichthyosis. |

What is ringworm ?

Skin infection in ringworm is caused by different dermatophytes (fungi) like *Trichophyton*, *Epidermophyton* or *Microsporum*. The lesion may be single or multiple, small or large, usually circular (ring-like, hence known as ringworm) with papulo-vesicular scaly active border, itchy with central clearing, spread peripherally and heals with hyperpigmentation. The prefix *tinea* (which indicates ringworm) follows the affected region of the body like *tinea corporis* (body), *tinea cruris* (thigh and buttocks), *tinea capitis* (scalp), *tinea pedis* (feet), *tinea manuum* (hand), *tinea barbae* (beard) and *tinea unguium* (nail).

Diagnosis is done by demonstration of hyphae on examination of skin scrapings, hairs or nail clippings after dissolving in a drop of 10-20% KOH solution for several hours, and examined under light microscope. Treatment is done by local application of antifungal agents (miconazole, econazole, clotrimazole, tolnaftate, Whitfield ointment etc); lotions and powders are preferred for intertriginous and hairy (scalp) areas. Systemic antifungal antibiotic like griseofulvin (10 mg/kg/day, after meals in twice daily dose for several weeks) is used in extensive lesion and in affection of scalp, nail and bearded area. Griseofulvin-resistant cases may be treated by oral ketokonazole (200 mg once or twice daily). Itraconazole (100-200 mg/day), fluconazole (150 mg/week) or terbinafine (250 mg/day) orally may be given for 4-8 weeks.

* *Tinea pedis* = athlete's foot, and *tinea cruris* = 'Dhobi' itch.

Treatment of pityriasis versicolor :

1. Maintenance of good personal hygiene.
2. Sodium thiosulphate (25% aqueous solution) once daily for 3 weeks used locally.

3. Selenium sulphide (2-5%) Apply at night and wash-off in the following morning; for 4 weeks.
4. Topical tolnaftate (1%), clotrimazole, miconazole (2%), econazole, ketoconazole (2%) may be tried — Apply twice daily for 10 days.
5. Retinoic acid (0.025%) cream may be of some help.
6. Azoles Oral ketoconazole 200 mg OD/BD, or oral itraconazole 100 mg, BD is used for 1 week in resistant cases. Ketoconazole shampoo may be tried.

Conclusion :

Mention the finding of pityriasis versicolor in long/short case, if present.

Case 59

HAEMORRHAGIC SPOTS

What are these ?

These are red, blue or black spots in the skin deep to the epidermis. As they contain extravasated blood, they do not disappear on application of pressure by a pinhead, glass slide (diascopy) or on stretching the skin of the affected areas.

How to classify haemorrhagic spots ?

The arbitrary classification goes like :

1. **Petechiae** - 1-2 mm in diameter (i.e., pinhead-size, punctate or tiny).
2. **Purpura** - a bit larger, 2-5 mm in diameter.
3. **Ecchymosis (bruise)** - large confluent (> 5 mm) purpuric lesions.
4. **Haematoma** - large haemorrhages in the skin with surface elevation (fluctuant collection).

Petechiae and purpura are collectively known as '**purpuric spots**'. Day by day, there is progressive colour changes in haemorrhagic spots.

Differential diagnosis :

1. Telangiectasia - small, dilated blood vessels visible on skin surface, specially on the lips.
2. Mosquito bite marks.
3. Spider naevi.
4. Cherry angioma or Campbell de Morgan's spot.
5. Drug rash (H/O offending drug intake, often blanches, may be itchy).

How to differentiate haemorrhagic spots from others ?

Telangiectasia, spiders, fresh mosquito bite marks blanch on application of pressure given by a pinhead or glass slide, and the colour returns on releasing the pressure. **Haemorrhagic spots never blanch on compression (identification point).**

Significance of haemorrhagic spots :

Haemorrhagic spots may be due to (A) and/or (B) and/or (C) mentioned below :

- (A) Thrombocytopenia (quantitative defect)— Either due to diminished production or increased destruction of platelet (e.g., idiopathic thrombocytopenic purpura or immune thrombocytopenic purpura [ITP]).
- (B) Vessel wall abnormalities, vasculitis or increased capillary fragility— Seen in Henoch-Schonlein purpura (HSP), scurvy, uraemia, ageing etc.
- (C) Platelet functional (qualitative) defect—Thrombasthenia or Glanzmann's disease (defect in platelet aggregation), after use of drugs like aspirin or phenylbutazone.

Causes of non-thrombocytopenic purpura :

- (A) Vessel wall abnormalities, e.g.,
 1. Infection— Meningococcus, viral infection, septicaemia.
 2. Henoch-Schonlein purpura (HSP) or allergic purpura.
 3. Hereditary haemorrhagic telangiectasis (HHT).
 4. Drugs— Penicillin, sulphonamides, corticosteroids.

5. Senile purpura.
6. Purpura simplex (devil's pinches).
7. Cushing's syndrome.
8. Scurvy.
9. Paraproteinaemias.
10. Vasculitis.
11. Uraemia.
12. Purpura fulminans.
13. 'Easy bruising syndrome' (benign disorder)

(B) Platelet functional defects (defects in platelet adhesion, aggregation or granule release).

* In (A) : BT — Normal or increased; platelet count — Normal.

In (B) : BT — Increased; platelet count — Normal.

Diagnosis of thrombocytopenia from history :

Apart from easy bruisability and haemorrhagic spots in the skin (petechiae, purpura or ecchymosis), the patient gives H/O spontaneous bleeding from nose (epistaxis) and gums (mucous membrane), haematuria, melaena or menorrhagia.

Importance of splenomegaly with purpuric spots :

1. Acute leukaemias.
2. SLE, SBE.
3. Lymphomas.
4. Blast crisis of CML and CLL.
5. ITP (10% cases only)—splenomegaly does not favour the diagnosis of ITP.
6. Myelofibrosis.
7. Hypersplenism due to any cause.

Laboratory tests of ITP :

1. Low total platelet count (normal platelet count is 1.5 lacs-4 lacs/ mm³). Peripheral blood smear may show large platelets.
2. Prolonged bleeding time (BT) with normal coagulation time (CT).
3. Antiplatelet antibodies may be found in blood.
4. To evaluate the secondary causes of ITP—serology for hepatitis C, HIV infection and ANF for SLE are done.
5. Bone marrow examination reveals megakaryocytes in normal or increased numbers with normal morphology in a hypercellular marrow. Sometimes, there are increased vacuolisation and diminished number of granules in the megakaryocytes.

* Normal BT is 2.5-10 minutes (Ivy) and normal CT is 9-15 minutes (glass tubes).

** Bleeding time (BT) is a sensitive measure of platelet number as well as function.

Characteristics of HSP (anaphylactoid purpura) :

1. A self-limiting vasculitis which often occurs after upper respiratory tract infection. It usually affects children and young adults.
2. The purpuric rashes are urticarial (palpable and itchy) and at times bilaterally symmetrical.
3. The rashes are usually present in buttocks, extensor surfaces of arms and lower legs; face and trunk spared—some purpuric rash coalesce and become necrotic.
4. HSP is often associated with haematuria and proteinuria (focal segmental proliferative glomerulonephritis), arthralgia or arthritis, and colicky abdominal pain.
5. The coagulation profile (BT and CT) remains normal.

* Thrombocytopenia = flat purpura, and vasculitis = raised purpura.

Probable causes of palpable purpura :

(A) Vasculitis due to any cause—HSP, polyarteritis nodosa, leucocytoclastic vasculitis etc.

(B) Infective emboli—meningococcaemia, disseminated gonococcal infection, measles, septicaemia.

* Palpable purpura are raised from surface, present in buttocks and extensor surfaces, and often pruritic.

Demonstration of Hess' capillary fragility test :

It is a test for diagnosis of increased capillary fragility (e.g., dengue fever) or immune thrombocytopenic purpura (ITP).

y

The procedure goes like this -

1. Blood pressure cuff is wrapped in upper arm and pressure is raised. The pressure is maintained halfway between systolic and diastolic BP for 5 minutes.
2. A circle of 1" diameter is drawn on the anterior aspect of upper forearm. The haemorrhagic spots already present within the circle are noted.
3. Normally after application of pressure by BP cuff, upto 10 new haemorrhagic spots may appear. But in pathological states (mentioned above), more than 20 new spots (diagnostic) may appear within the circle after deflation of BP cuff.

This test may be negative in mild to moderate thrombocytopenia but is positive in severe thrombocytopenia (ITP). This test is also known as TOURNIQUET TEST.

Cause of sudden death in thrombocytopenia :

Sudden massive haemorrhage in any vital organ like intracerebral haemorrhage may be fatal in a patient of thrombocytopenia.

Thrombocytosis and thrombocythemia :

- (A) Thrombocytosis — Platelet > 4 lacs / mm³ which is a temporary elevation e.g., splenectomy, severe bleeding etc. Thrombocytosis is regarded as part of acute phase reaction.
- (B) Thrombocythemia — Platelet > 8 lacs / mm³ which is a sustained elevation. It is regarded as a myeloproliferative disorder.

* Platelet count < 1 lac / mm³ is known as thrombocytopenia.

** 1 unit of platelet transfusion raises the count by 10000/mm³.

Purpura in young and elderly :

- (A) Young ITP, HSP, SLE, drug-induced, acute leukaemias, infections (e.g., meningococcus).
- (B) Elderly—Senile purpura, leukaemia, drug-induced, scurvy, paraproteinaemias.

Drugs causing thrombocytopenia :

1. Sulphonamides.
2. Penicillin.
3. Cyclophosphamide.
4. Thiazides.
5. Quinine.
6. Carbamazepine.
7. Aspirin.
8. Heparin.

Common medical causes of purpura :

1. ITP
2. Acute leukaemias.
3. Aplastic anaemia (no splenomegaly).
4. Drugs (no splenomegaly).
5. SLE
6. Lymphoma.
7. Hepato-cellular failure.
8. SBE

Causes of gum bleeding :

1. Gingivitis.
2. ITP.
3. Acute leukaemias.
4. Aplastic anaemia.
5. Scurvy.
6. Hemophilia.
7. Anticoagulant therapy.
8. Vincents infection.

Causes of gum hypertrophy :

1. Poor oral hygiene.
2. Phenytoin therapy in epileptics.
3. Pregnancy
4. Scurvy
5. Acute monocytic leukaemia.
6. Idiopathic familial fibromatosis.
7. Nifedipine or amlodipine therapy
8. Histiocytosis X disease.

Medical causes of epistaxis :

1. Systemic hypertension.
2. Bleeding disorders (ITP, haemophilia)/anticoagulant therapy/DIC; acute leukaemias.
3. Vasculitis.
4. High altitude.

5. Rendu-Weber-Osler disease (hereditary haemorrhagic telangiectasis).
6. Uraemia.
7. Kala-azar, malaria, enteric fever, infectious mononucleosis, rheumatic fever, diphtheria.

Purpuric eruption with pyrexia :

- | | |
|------------------------------|---------------------------------------|
| 1. Dengue. | 6. Gram-negative septicaemia. |
| 2. Infective endocarditis. | 7. Rocky Mountain spotted fever. |
| 3. Meningococcaemia. | 8. Disseminated gonococcal infection. |
| 4. Purpura fulminans. | 9. Cutaneous small vessel vasculitis. |
| 5. Viral haemorrhagic fever. | 10. Rat-bite fever. |

Safe analgesics in purpura :

Codeine and paracetamol have no effect on platelet number and function.

Critical platelet count :

It is 20000/ mm³. Below this level, spontaneous haemorrhage in any vital organ (e.g., intracerebral haemorrhage) or anywhere in the body may endanger the life of the patient. These patients require immediate temporary support with platelet or fresh blood transfusion. Recently, haematologists opined that the level may be near about 10000/mm³ to have risk of internal bleeding.

Sites and organs to be examined in a case of purpura :

Read the 'Scheme of examination' from 'Lymphoreticular system'.

What do you mean by 'bleeding disorders' ?

Bleeding disorders are usually due to one of the following abnormalities :

- a) Coagulation disorders.
- b) Platelet disorders (number and / or function defect), and
- c) Vessel wall abnormalities.

A detailed history, meticulous physical examination and few initial screening tests are required to evaluate bleeding disorders.

- (A) **Coagulation disorders** (e.g., haemophilia, Christmas disease) : Only in males (haemophilia), family history +ve (due to X-linked inheritance, there is oblique transmission, i.e., uncles and nephews may be the sufferer), life-long history; bleeding into viscera, retroperitoneum, muscles (calf, psoas, tongue) and joints (e.g., knee haemarthrosis) are characteristic; purpura is rare; bleeding from nose, mouth, G. I. tract, urinary tract, wounds or in the subcutaneous tissue may occur. Bleeding starts several hours after trauma or surgery (H/O induced and prolonged bleeding), and is notorious for persistence (not profuseness); local pressure is ineffective; absence of splenomegaly, normal BT with prolonged CT, while platelet count remains normal. Acquired coagulation disorder may result from vitamin K deficiency or oral anticoagulant therapy.
- (B) **Platelet disorders** : H/O drug intake (specially, aspirin), and bleeding occurs characteristically from mucous membrane (gum), nose (epistaxis), skin (petechiae, purpura, bruises), buccal mucosa, and per vaginum; bleeding may occur from G. I. tract, CNS or superficial wounds; bleeding happens to occur immediately after trauma or surgery, and local pressure is effective to stop bleeding; may have splenomegaly, increased BT with normal CT, while low platelets in thrombocytopenia and normal platelets in thrombasthenia. Qualitative disorders of platelet are adhesion defect (von Willebrand's disease, Bernard-Soulier syndrome) and aggregation defect (Glanzmann's thrombasthenia, uraemia, aspirin therapy).
- (C) **Vessel wall abnormalities** : Consider age of the patient (common in old age), petechiae/purpura, recurrent bleeding at a single site, bleeding occurs immediately after trauma or surgery, and local pressure is effective. There is absence of splenomegaly, normal or elevated BT with normal CT, and normal platelet count. See page 368 for causes of vessel wall abnormalities.

* (B) and (C) are commonly characterised by easy bruising and spontaneous bleeding.

Conclusion :

In the presence of purpuric spots, one should examine the patient for 'blanching reaction' which is always absent here. Always perform the Hess' capillary fragility test in a suspected patient of ITP.

Case 60

LEPROSY

Definition :

This is a chronic granulomatous disease caused by *Mycobacterium leprae* (a gram positive, acid- and alcohol-fast bacillus) which principally attacks the skin, peripheral nerves and nasal mucosa. It is also known as **Hansen's disease**.

What are the different forms ?

1. Two polar forms are—
 - (i) Lepromatous (LL), and
 - (ii) Tuberculoid (TT).
2. Borderline or dimorphous leprosy—
 - (i) Borderline tuberculoid (BT).
 - (ii) Borderline (BB).
 - (iii) Borderline lepromatous (BL).
3. Indeterminate leprosy (I)—
 - (i) Non-classifiable early lesion.
 - (ii) Transforms into any polar form, or
 - (iii) May spontaneously remit.

* Principally, there are two main forms : polar forms and borderline leprosy.

What do you mean by LL and TT forms ? Multibacillary or paucibacillary ?

Lepromatous leprosy (LL) means :

- (i) Highly bacillated (multibacillary),
- (ii) Low resistance (impaired cell-mediated immunity), and
- (iii) Systemic infectious form.

Tuberculoid leprosy (TT) means :

- (i) Non-infectious (paucibacillary),
- (ii) High resistance (i.e., high cell-mediated immunity), and
- (iii) Localised disease.

(A) Multibacillary : BB, BL and LL.

(B) Paucibacillary : I, BT and TT.

* Multibacillary—has 6 or more skin lesions which may have bacilli.

Paucibacillary—has 5 or less skin lesions with no bacilli.

Describe the skin lesion of TT variety :

1. Solitary lesion or a couple of lesions.
2. Macular or raised as plaques, or as rings with flat centre (central healing).
3. Sharply defined or circumscribed.
4. *Hypopigmented*. dry, scaly lesion.
5. *Hypoaesthetic*, hypohydrotic (diminished sweating) and hypotrichotic (loss of hair).
6. Present anywhere in the body specially in arms, buttocks, legs and face.

PLUS

The nerve twig or the peripheral nerve supplying the skin lesion may be thickened, e.g.,

- a) Great auricular nerve across the sternomastoid muscle (in the posterior triangle) in neck.
- b) Ulnar nerve at elbow.
- c) Median nerve at wrist.
- d) Radial cutaneous nerve at wrist.
- e) Common peroneal nerve at the neck of fibula.
- f) Posterior tibial nerve around medial malleolus.
- g) Sural nerves at the lateral part of ankle.

N.B. : It is compulsory to test the touch sensation in the skin lesion in all suspected patient of TT variety. Always palpate the nerves proximal to the skin lesion. One should look for the great auricular nerve in the neck (by turning the head to the opposite side, the nerve stands out), the ulnar nerve in the funny bone or the common peroneal nerve at the neck of fibula. Test the nerves for thickening, tenderness, and its motor as well as sensory functions.

Describe the skin lesion of LL variety :

1. Hypopigmented macules, plaques or *nodules* (may be bilaterally symmetrical) widely scattered in the body. *Gradually the skin becomes infiltrated and thickened.*

2. Ill-defined (with margins that merge imperceptibly with normal skin).
3. The surface of the lesion is smooth, shiny but without any central healing (inverted-saucer).
4. If the lesion advances, 'leonine' facies may be seen.
5. Both the loss of cutaneous sensation and nerve involvement are late features.

PLUS

- a) Lymphadenopathy (painless).
- b) Hepatosplenomegaly.
- c) Peripheral neuropathy with trophic changes (watch the sole of the feet for trophic ulcer).
- d) Testicular atrophy (with impotence) and gynaecomastia; pedal oedema.
- e) Ocular lesions — Keratitis, iridocyclitis etc.
- f) Miscellaneous—anaemia, nephrotic syndrome, nasal collapse.

N.B. : In a suspected patient of LL variety, one should examine the eyes (e.g., corneal anaesthesia, corneal ulcer), palpate the liver, spleen and testes, test for peripheral neuropathy, search for lymphadenopathy and examine for gynaecomastia. Look at the face carefully and never forget to test the touch sensation in the skin lesion. Palpate the nerves proximal to the skin lesion (though a late and non-prominent feature).

Different types of nerve involvement in leprosy :

- (A) Nerver thickening—Classical description given in TT variety (see above).
- (B) Sensory nerve involvement with gloves and stockings anaesthesia is commoner in LL variety (peripheral neuropathy).
- (C) Motor nerve involvement may result in muscle weakness, wasting, paralysis and contractures lately. The ulnar nerve (claw hand), median nerve (ape hand), common peroneal nerve (foot drop) or posterior tibial nerve (claw toes) may be affected; common in TT variety.
- (D) Cranial nerve involvement ; Facial (often bilateral) or trigeminal nerve (loss of corneal reflex); sometimes, multiple cranial nerves are involved (polyneuritis cranialis multiplex).

* Always consider leprosy as a possible cause of peripheral neuropathy in India. Central nervous system is not affected in leprosy.

** Ulnar nerve abscess with adjacent cellulitis in skin is common in BT variety.

Describe the leonine face :

- (A) Description ;
 1. Lines on the forehead become deeper as well as the upper central incisor teeth loosen or fall out and there is hoarse voice.
 2. Loss of hair in the lateral part of eyebrows (madarosis) and thick eyebrows.
 3. Depressed bridge of the nose, broad nose with nasal collapse (saddle nose).
 4. Thickened skin of face and forehead, specially the infiltrated earlobes.
 5. There may be perforated nasal septum.

- (B) Causes ;
 1. Leprosy (lepromatous).
 2. Albright's disease (fibrous dysplasia of bone).
 3. Carcinoid syndrome.
 4. Cleidocranial dysostosis.
 5. Primary hypertrophic osteoarthropathy (pachydermoperiostitis).
 6. Amyloidosis.
 7. B-cell lymphoma.
 8. Paget's disease.

* Actually 2 to 8 mimic leonine face.

** **Saddle nose** : lepromatous leprosy, ectodermal dysplasia and congenital syphilis; **destruction of nasal structures** : lepromatous leprosy, Wegener's granulomatosis, midline granuloma and lupus vulgaris.

D/D of multiple nodular lesions in face :

- | | |
|---|----------------------------|
| 1. Leprosy (lepromatous). | 4. Acne rosacea. |
| 2. Post Kala-azar dermal leishmaniasis (PKDL) | 5. Non-Hodgkin's lymphoma. |
| 3. Sarcoidosis. | 6. Amyloidosis. |

Do you know any other variety of leprosy ?

1. Histoid leprosy— Succulent cutaneous nodules and a variant of LL variety.
2. Neuritic leprosy— Peripheral nerves are thickened without any skin lesion, probably of TT type— common in India.

Describe the hand of a leprosy patient :

Read the section on 'Examination of the hands'.

Causes of thickened peripheral nerves :

- | | |
|--|---|
| 1. Leprosy. | 6. Sarcoidosis. |
| 2. Neurofibromatosis. | 7. Chronic Guillain-Barre syndrome. |
| 3. Acromegaly. | 8. Dejerine-Sottas type neuropathy. |
| 4. Amyloidosis (primary). | 9. Refsum's disease. |
| 5. Charcot-Marie-Tooth disease (juvenile). | 10. Injury to the nerve. |
| | 11. Idiopathic hypertrophic neuropathy. |

Complications of leprosy :

1. Crippling of hands.
2. Blindness (due to corneal anaesthesia).
3. Tuberculosis (one of the major cause of death).
4. Amyloidosis (secondary).
5. Leprosy reactions.
6. Social outcast.

Lepromin skin test in all the varieties of leprosy :

It depends on the cellular immunity or resistance of the patient though it has no value in diagnosing leprosy (as it may be positive in normal people). *It helps in classifying the disease.*

TT — Strongly positive.

LL — Negative.

BT — Positive.

BB — Negative or weakly positive.

BL — Negative. *

* In Lepromin test, suspension of dead bacilli (*M. leprae*) is injected intradermally, and the findings read after 4 weeks.

Reactions (lepra reactions) in leprosy :

- (A) Type I (reversal reaction)— Existing skin lesions may develop erythema and swelling, peripheral nerves become tender and painful, with sudden loss of nerve function (e.g., foot drop). Common in BL, BT and BB variety. It is a type IV delayed hypersensitivity reaction and is being treated by aspirin, corticosteroid and chloroquine.
- (B) Type II or erythema nodosum leprosum (ENL)— Common in LL or BL variety. It occurs in patients in their second year of treatment. Acute crops of painful, tender nodules develop with pain in the nerves, fever, arthritis, lymphadenopathy, iridocyclitis, bone pain and epididymo-orchitis. ENL is treated by analgesics, antipyretics, chloroquine, thalidomide (never used in pregnancy) or prednisolone, and increasing the dose of clofazimine. ENL develops due to immune complex-mediated vasculitis. Eye care is done by hydrocortisone or atropine drop.
- (C) Lucio phenomenon— Rare, occurs in diffuse non-nodular LL variety (develops angular ulcer).

Incubation period of leprosy and portal of entry of *M. leprae* :

Incubation period — Generally 2-6 years, may be more; 2-5 years for TT and 8-12 years for LL variety (as *M. leprae* has doubling time of 12 days i.e., very slow multiplication time).

Portal of entry— Through nasal mucosa or by inoculation through skin (needs prolonged close contact) Droplets from the sneezes of a lepromatous patient is highly infectious. Untreated patients (man is the only reservoir) with smear-positive extensive disease are potential source of infection.

What are the differential diagnosis of leprosy ?

- (A) Skin lesions -
 1. Hypopigmented lesions like vitiligo, psoriasis, pityriasis versicolor
 2. SLE.
 3. Lupus vulgaris (tuberculosis of skin).
 4. Yaws.

5. Dermal leishmaniasis.
6. Sarcoidosis.
- (B) Claw hand — Read the section on 'Claw hand'.
- (C) Peripheral neuropathy — Read the section on 'Peripheral neuropathy'.
- (D) Peripheral nerve thickening — Read the causes of 'thickened peripheral nerves' discussed above.

How to establish your diagnosis ?

The diagnosis is essentially clinical. The investigations performed are :

- (A) By slit-skin smear method — Principally the multibacillary disease (BB, BL and LL) is diagnosed by this method and it is less informative in paucibacillary (I, BT and TT) variety. The pinched skin between thumb and forefinger is incised by a scalpel and the exposed area is scraped. The juice is then stained by Ziehl-Neelsen's method (to find out acid-fast bacilli) and put under the microscope. Now the bacterial index (BI) i.e., the number of bacilli living or dead are calculated and scored on a logarithmic scale. Typical histology after skin biopsy also helps in diagnosis.

Sites of incision — Skin lesions (nodules), earlobes, dorsum of the middle or ring finger, nasal mucous membrane.

- (B) Biopsy of lymph nodes, nerves, liver may demonstrate the bacilli (rarely required).
 (C) Serology or PCR testing for *M. leprae*—neither sensitive nor specific for diagnosis.

Associated features — Mild anaemia, high ESR and hyperglobulinaemia (may give false-positive serological tests like VDRL, rheumatoid factor and ANF—in LL variety).

How are you going to treat a leprosy patient ?

Recommended treatment regimens in leprosy for adults (modified WHO guidelines)

(A) Paucibacillary disease (I, BT and TT) -

- (i) Rifampicin 600 mg — once monthly (single supervised dose).
 - (ii) Dapsone — 100 mg daily (self-administered dose).
- Duration — 6 months.

(B) Multibacillary disease (BB, BL and LL) -

- (i) Rifampicin — 600 mg once monthly (single supervised dose).
- (ii) Clofazimine — 300 mg once monthly (single supervised dose).

PLUS

- (iii) Daily dose of clofazimine (50 mg) and dapsone (100 mg)—both self-administered dose.
- Duration—12 months.

(C) Single lesion paucibacillary disease (I, BT and TT)-

- | | |
|--------------------------|--------------------|
| (i) Rifampicin 600 mg | } as a single dose |
| (ii) Ofloxacin 400 mg | |
| (iii) Minocycline 100 mg | |

* Bacteriostatic—dapsone and clofazimine: bactericidal—rifampicin.

** in pregnancy : clofazimine is not used, rifampicin is used cautiously in first trimester but dapsone is safe in pregnancy.

*** Because of long generation time of the bacillus, low dose rifampicin is sufficient (once monthly use).

**** Management of anaemia, secondary infection and lepra reaction are done accordingly.

Side-effects of clofazimine and dapsone :

(A) Clofazimine -

- | | |
|--|-----------------------------------|
| 1. Skin pigmentation (red colouration of skin, urine and body secretions). | 3. Pruritus and ichthyosis. |
| 2. Abdominal pain. | 4. Anorexia, nausea and vomiting. |

(B) Dapsone (diaminodiphenyl sulphone / DDS)—

- | | |
|----------------------------|------------------------------------|
| 1. Exfoliative dermatitis. | 5. Avoided in G_6 PD deficiency. |
| 2. Haemolytic anaemia. | 6. Psychosis. |
| 3. Agranulocytosis. | 7. Hepatitis. |
| 4. Methaemoglobinaemia. | 8. Hypoproteinaemia. |

Use of rifampicin in internal medicine :

1. Tuberculosis and in non-tuberculous mycobacteria.

2. Leprosy.
3. Chemoprophylaxis of persons at risk of developing meningococcal meningitis.
4. Legionnaires' disease.
5. Brucellosis (with co-trimoxazole or tetracycline-streptomycin combination).
6. Eradication of chronic carrier state in enteric fever (with co-trimoxazole).
7. Endocarditis as a result of *Staphylococcus epidermidis* or *Corynebacterium Spp.* (often with vancomycin).
8. Anthrax.
9. Anaplasmosis.
10. Primary amoebic meningoencephalitis (caused by *Naegleria fowleri*).
11. Pruritus (may be used in pruritus associated with cholestasis).
12. Bartonellosis (chronic).
13. Cat-scratch disease.
14. Prevention of *Haemophilus influenzae*.

Other drugs used in leprosy :

- | | |
|------------------------|---|
| 1. Acedapsone (DADDs). | 6. Minocycline. |
| 2. Ethionamide. | 7. Ofloxacin. |
| 3. Thiacetazone. | 8. Clarithromycin. |
| 4. Prothionamide. | 9. Streptomycin / kanamycin. |
| 5. Thiambutosine. | 10. Corticosteroid (in lepra reactions) |

* Vaccination by BCG with heat-killed *M. leprae* is being tried for prevention of leprosy.

Other modalities of treatment in leprosy :

1. Paralysis from reactional neuritis— Splinting of the limb.
2. Tarsorrhaphy for corneal anaesthesia.
3. Trophic ulcer in feet— Rest, use of crutches, walking plaster.
4. Infection— Appropriate antibiotic.
5. Special protective footwear for anaesthetic feet.
6. Plastic surgery in face or reconstructive surgery in limbs.
7. Haematinics and multivitamins.
8. Physiotherapy to prevent contracture and muscle atrophy.
9. Rehabilitation.

* 'Hypopigmentation' in leprosy is due to 1. Direct invasion at melanocyte level, 2. Utilisation of dopa by lepra enzyme system, and 3. Vascular changes leading to atrophy of melanocytes.

Case 61

NEUROFIBROMATOSIS

What is neurofibroma ?

These are benign, soft, usually non-tender papules or nodules varying in size from a pea to a cricket ball which develops from the Schwann cells and fibroblasts of the neurilemmal sheath of the peripheral nerve (a form of hamartoma). Majority are symptomless but occasionally there is paraesthesia. **The mass may be moved from side to side only** but is otherwise fixed by the nerve from which it arises.

What is neurofibromatosis ?

It is also known as "von Recklinghausen's disease" and has an autosomal dominant inheritance. Here, multiple neurofibroma are associated with cafe-au-lait spots and axillary freckling.

Cafe-au-lait (colour of white coffee or coffee in milk) spots are round to ovoid, pale yellow-brown macules that vary in diameter from 1 cm to more than 15 cm and is usually present on the trunk.

* Friedrich Daniel von Recklinghausen (1833-1910) was Professor of Pathology, Strasbourg, France.

Conditions associated with cafe-au-lait spots :

1. Neurofibromatosis which is characterised by,
 - (i) Six or more spots.
 - (ii) Uniformly hypomelanotic, circumscribed.
 - (iii) Diameter >1.5 cm with multiple Lisch nodules in iris (1 mm yellow-brown spots in iris, best revealed by slit-lamp examination).
2. Albright's disease (polyostotic fibrous dysplasia)—larger and more irregular than No. 1.
3. Occasionally in normal population.
4. Multiple endocrine adenomatosis-type III.
5. Tuberous sclerosis.
6. Watson's syndrome (neurofibroma with pulmonary stenosis).
7. Ataxia-telangiectasia.
8. Bloom syndrome.

What are the types of neurofibromatosis ?

- (A) Type I (von Recklinghausen's disease) — Peripheral form (found in 70% cases) and thus, readily recognized by the cutaneous lesions. It is due to an abnormal gene carried on chromosome 17 which encodes 'neurofibromin'. This type may be associated with scoliosis, plexiform neurofibroma, aqueduct stenosis. Lisch nodules in iris and rarely pheochromocytoma.
- (B) Type II — Central form and difficult to diagnose as there are few or no cutaneous lesion. This type may be associated with optic glioma, acoustic neuromas, meningiomas, entrapment of spinal nerve roots, cataract, mental retardation and epilepsy. The gene for this type is located on chromosome 22 which encodes 'Merlin' or Schwannomin. As this type may have no cutaneous stigmata, a family history of cerebral or spinal lesion should be noted very carefully.

Where the neurofibromas are mostly found ?

Mainly they are present by the sides of the neck and in the extremities (cafe-au-lait spots are primarily present on the trunk). The tumours are felt as movable, bead-like nodules. Rarely, they may be painful and tender on pressure.

When to investigate neurofibromatosis ?

In the presence of :

1. Symptoms of cerebral or spinal involvement (acoustic neuroma may present as cerebello-pon-tine angle tumour, and spinal involvement may present as paraplegia), and
2. Malignant change (sarcomatous change in very few cases).

What is plexiform neurofibroma ?

This is diffuse neurofibromatosis with an overgrowth of the skin and subcutaneous tissue. In this way large hanging folds of skin may be formed. The commonest sites are temple (in connection with branches of trigeminal nerve), the upper eyelid and the back of the neck. The severe form of this variety is known as 'elephantiasis neuromatosa' (usually involves the lower limb).

What is pachydermatocele ?

It is a very rare variety of neurofibromatosis in which coils of soft tissues hang around the neck.

What is phakomatosis ?

These are a unique group of diseases in which neurological abnormalities are combined with congenital defects in skin, retina and other organs. The causes of phakomatosis are von Recklinghausen's disease, tuberous sclerosis, von Hippel-Lindau syndrome and Sturge-Weber disease. These are also known as 'congenital neurocutaneous syndrome'.

'Phakoma' (collection of abnormal neuroglial tissue) is seen in the retina (greyish circular mass about the size of half the optic disc) by ophthalmoscopy.

Conclusion :

If you see a patient with neurofibroma, always search for —

1. Scoliosis, 2. Cafe-au-lait spots, and 3. Axillary freckles,

* Biopsy is usually contraindicated in neurofibroma as it may change into neurofibrosarcoma.

** Surgery may be considered for cosmetic reason or pressure symptoms.

Case 62

Scleroderma

Describe the face of the patient :

The term scleroderma (henceforth called systemic sclerosis—SSc) refers to thickening or hardening of skin due to abnormal collagen deposition in dermis. The typical 'facies' goes like this—

1. *Mask-like facies* (loss of facial expression).
2. Absence of normal skin wrinkling ('ironed' out skin folds).
3. Pinched-up nose or beaking of the nose.
4. Inability to open the mouth fully — Small mouth (*microstomia*) with radial furrows on closing (tobacco-pouch, fish-mouth or crow-feet appearance).
5. Skin over the face seems taut and shiny.
6. Pigmentation and depigmentation.
7. Telangiectasia over face and lips.

Common causes of masked facies :

Poverty of expression in face (hypomimia) is seen in,

1. Parkinsonism.
2. Scleroderma.
3. Hypothyroidism (myxoedema).
4. Bilateral UMN type of facial nerve palsy.
5. Depression.
6. Sometimes in myasthenia gravis and facial myopathies.
7. Dementia.
8. Sometimes in pseudobulbar palsy.

Types and subtypes :

- (A) Systemic sclerosis (SSc) is a multisystem disorder of unknown aetiology affecting skin (*scleroderma*), CVS, G.I. tract, kidneys, musculo-skeletal system, lungs and vasculature. Localised form without systemic involvement is known as *localised scleroderma*.
- (B) SSc is divided into two subgroups : *limited cutaneous scleroderma* (LcSSc; **70%**) and *diffuse cutaneous scleroderma* (DcSSc; **30%**). LcSSc → skin involvement over sites restricted distal to elbow or knee. Face is always involved. LcSSc is now synonymous with CREST syndrome (see afterwards), and DcSSc → involvement of skin of fingers rapidly involves the whole body, trunk and face.
- (C) Localised scleroderma is again divided into two subgroups : *morphea* and *linear scleroderma*.
- (D) *Scleroderma sine scleroderma* : Systemic features without any skin involvement.

Initial suspicion of scleroderma in a patient :

1. While examining the patient for anaemia i.e., when the lower eyelids are retracted downward, it seems very tight and the lower palpebral conjunctiva is not exposed fully.
2. Characteristic facies, specially in a middle-aged female patient (30-50 years).

Stages of scleroderma :

There are three stages —

1. Oedematous stage (non-pitting oedema).
2. Indurated stage.
3. Atrophic stage.

How the patients present themselves ?

Very often the first symptom is Raynaud's phenomenon, or symmetrical swelling or stiffness of the fingers.

Examine the hand of your patient (scleroderma) :

1. Skin is very taut, firm and thick, and the skin of the fingers cannot be pinched-up (sclerodactyly) as it is tightly bound to underlying subcutaneous tissue.
2. Raynaud's phenomenon.

3. Symmetrical swelling (puffiness) of fingers.
4. Limitation of full extension and flexion of fingers (flexion contracture).
5. Pulp atrophy (soft tissue at fingertips is lost).
6. Skin is dry, coarse with loss of hair; skin may appear shiny.
7. Skin may be pigmented and/or depigmented (salt-pepper appearance); telangiectasia may be seen.
8. Nail-fold thrombi, dilated nail-fold capillary loops, digital infarcts, painful digital ulceration and rarely digital gangrene.
9. Pseudoclubbing may be present (due to resorption of bone of terminal phalanx).
10. Calcific deposits (calcinosis cutis) in the subcutaneous tissue (sometimes skin may break down with discharge of calcific material).

* The changes in the nail-fold capillaries are best examined clinically by an ophthalmoscope or capillary microscope.

Causes of deformed/mutilated fingers/toes :

- | | |
|-----------------------|------------------------------------|
| 1. Leprosy. | 9. Atherothrombosis. |
| 2. Scleroderma. | 10. Antiphospholipid syndrome. |
| 3. Buerger's disease. | 11. Arthritis mutilans (psoriasis) |
| 4. Congenital defect. | 12. Trauma. |
| 5. Frost bite. | 13. Porphyria. |
| 6. Diabetic foot. | 14. Amyloid neuropathy. |
| 7. Syringomyelia. | 15. Lesch-Nyhan syndrome. |
| 8. Vasculitis. | |

Respiratory system involvement in SSc :

Respiratory system involvement is the major cause of morbidity and mortality in SSc.

1. Aspiration pneumonia (due to dysphagia).
2. Interstitial lung disease ('honeycomb lung' in chest X-ray).
3. Pulmonary arterial hypertension (10-15%)—common in long-standing disease.
4. Chronic cor pulmonale.
5. Malignant alveolar or bronchiolar cell neoplasm.
6. Pleurisy, rarely.
7. Restriction of chest movement (due to very tight skin which looks like Roman breast plate)—Hidebound chest.

CVS involvement in SSc :

1. Pericarditis.
2. Heart block.
3. Arrhythmia.
4. Restrictive cardiomyopathy (myocardial fibrosis).
5. Heart failure (right heart failure from cor pulmonale, and left heart failure from cardiomyopathy).
6. Systemic hypertension.

Gastrointestinal tract involvement in SSc :

1. Heartburn, regurgitation of gastric contents due to reflux oesophagitis (symptoms are more when the patient lies flat).
2. Dilatation and atony of distal oesophagus — peptic oesophagitis.
3. Dysphagia, specially to solid foods (manometry shows decreased amplitude or disappearance of peristaltic waves in lower 2/3rd of the oesophagus) due to oesophageal hypomotility; odynophagia may occur.
4. Delayed gastric emptying due to atony and dilatation.
5. Recurrent occult upper G. I. bleeding indicates 'watermelon stomach' i.e., antral vascular ectasia.
6. Malabsorption syndrome (due to bacterial overgrowth).
7. Chronic constipation, distension and sometimes intestinal pseudo-obstruction (due to atony).
8. Pneumatosis cystoides intestinalis (radiolucent cysts within the wall of small intestine).

* Dysphagia is a very important symptom of SSc.

Musculo skeletal manifestations of SSc :

1. Symmetrical polyarthritis (like rheumatoid arthritis).
2. Restriction of hand movement (mostly due to stiffness of skin).
3. Leathery crepitations can be palpated over tendon sheaths or moving joints (specially knee joints) in an advanced disease.
4. Carpal tunnel syndrome.
5. Resorption of terminal phalanges resulting in pseudoclubbing.
6. Proximal muscle weakness, wasting (due to disuse) and low grade myositis.

Renal involvement in SSc :

1. Hypertension.
2. Non-progressive proteinuria.
3. 'Scleroderma renal crisis'—it is due to obliterative vasculopathy of renal cortical arteries and is one of the common cause of death.

How to assess the severity of scleroderma at the bedside ?

Normally in health, a person's 4 fingers (excluding thumb) easily enter within his oral cavity when placed side by side. In scleroderma, as the mouth becomes smaller, all the four fingers may not enter within the oral cavity. Observe the number of patient's fingers entering within the oral cavity with ease; if it is 3 fingers, it is grade I and if it is 1 finger, it is of grade III (severe).

What are the differential diagnosis ?

1. Causes of Raynaud's phenomenon (see below).
2. CREST syndrome (calcinosis, Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly and telangiectasia). This is synonymous or a subset of limited cutaneous scleroderma.
3. Eosinophilic fascitis.
4. Pseudo-scleroderma (amyloidosis, scleredema, acromegaly and scleromyxoedema).

Drugs or toxins producing scleroderma like lesion :

- | | |
|--------------------|------------------------------------|
| 1. Vinyl chloride. | 3. Bleomycin. |
| 2. Pentazocine. | 4. Epoxy and aromatic hydrocarbon. |

How to confirm your diagnosis ?

Antinuclear factor (ANF) is present in 95% cases. Antinuclear antibodies to anti-topoisomerase I (anti-Scl-70) is found in 30% patients of diffuse cutaneous variety; anticentromere antibodies are found in 75% cases of limited cutaneous variety (e.g., CREST syndrome). Rheumatoid factor is positive in 30%.

Chest X-ray (to see cardiomegaly), X-ray of hands (acrosteolysis or calcinosis), barium swallow (confirms oesophageal hypomotility) and CT scan of thorax (to demonstrate interstitial lung disease) also help in diagnosis. Skin biopsy is not so important in diagnosis.

What is mixed connective tissue disease (MCTD) ?

It is an overlap syndrome characterised by the combination of SLE, scleroderma, polymyositis and rheumatoid arthritis. It is associated with unusually high titres of circulating antibody to a nuclear ribonucleoprotein (U1-RNP).

What is Raynaud's phenomenon ?

Definition — It is a vasospastic disorder (intense vasospasm of peripheral arteries), manifested clinically by the classical 'triphasic colour response' which is sequential development of digital blanching (pallor due to vasospasm of arteries), cyanosis (blue due to sluggish blood flow and accumulation of deoxygenated blood following ischaemia) and rubor (redness due to vasodilatation and reactive hyperaemia) of the fingers and toes following cold exposure and subsequent rewarming. The changes in the fingers and toes are often diagnosed by nail-fold capillography. The fingers are affected more than toes. Numbness, burning and severe pain in digits are common features.

It is a cold-induced digital ischaemia secondary to reflex sympathetic vasoconstriction. The duration of the attack is variable and may last for hours. Some patients may experience bicolour response (e.g., pallor and cyanosis). In chronic and severe cases, there may be tissue infarction and loss of digits.

Common causes are :

1. Collagen vascular diseases like scleroderma, dermatomyositis, CREST syndrome, SLE etc.
2. Raynaud's disease (women of late teens, bilateral and symmetrical affection of hands).

3. Throacic inlet syndrome (specially in unilateral involvement).
4. Neurological — Syringomyelia, spinal cord tumours.
5. Trauma — Vibration injury, use of pneumatic drills, cold injury, typing.
6. Blood dyscrasias — Myeloproliferative disorders, hyperviscosity syndrome, cryoglobulinaemia.
7. Drugs — Ergot derivatives, (3-blockers, methysergide).
8. Arterial disease— When affected by microemboli e.g., atherosclerosis, Buerger's disease.

N.B. : Now-a-days Raynaud's phenomenon is broadly divided into two categories :

- a) The idiopathic variety called Raynaud's disease (No. 2), and
- b) The secondary variety described above (No. 1 to 8 except No. 2).

* D/D of Raynaud's phenomenon are acrocyanosis, erythromelalgia, chilblain, cold agglutinin disease.

D/D of hardness (or thickening) of skin :

1. Morphea (localised scleroderma)—presents as solitary or multiple patches of thickened skin.
2. Lymphoedema.
3. Vascular insufficiencies (chronic).
4. Recurrent cellulitis with venous stasis.
5. Lipoid proteinosis.
6. Carcinoid syndrome.
7. Schirrhous carcinoma (hardening of skin of chest).
8. Porphyria cutanea tarda.
9. Chronic graft-versus-host disease.
10. Toxic oil syndrome (linked to consumption of contaminated rapeseed oil).
11. Eosinophilia-myalgia syndrome (due to consumption of L-tryptophan).
12. Nephrogenic systemic fibrosis (in uraemic patients on haemodialysis).

Treatment modalities used in SSc :

Virtually no agent has been shown to arrest skin changes. The drugs used are :

1. D-penicillamine (diminishes collagen cross linkage and reduce skin thickening).
2. Colchicine (diminishes procollagen to collagen conversion).
3. Treatment of reflux oesophagitis (by PPI and prokinetics), myositis (by glucocorticoids), articular symptoms (by NSAID), respiratory tract infection (by antibiotics), interstitial lung disease (by glucocorticoids and immunosuppressants), CVS involvement (pericarditis by glucocorticoids) and hypertension (ACE inhibitors) due to renal involvement are done accordingly.
4. 'Scleroderma renal crisis' is a medical emergency, and requires intensive control of hypertension and use of ACE inhibitors.
5. IFN- α , IFN- γ and human recombinant relaxin are being tried with mixed results.
6. Clinical trials of immunosuppressive drugs (methotrexate, mycophenolate mofetil, cyclosporine, azathioprine) are underway.
7. **Raynaud's phenomenon** : patients should wear gloves and woolen mittens, minimise cold exposure, protect the body with warm clothings and abstain from tobacco smoking. Drugs used are calcium channel blockers (nifedipine or diltiazem), prazosin, losartan, sildenafil, topical nitroglycerine, I.V prostaglandins, aspirin/dipyridamole (to prevent platelet aggregation), endothelin-1 receptor antagonist (bosentan, specially in digital ulcers; costly), calcitonin gene-related peptides and statins (empirical use). One should avoid (3-blockers, ergotamine, sympathomimetics and OC pills. Digital sympathectomy may be effective in some patients.
8. Regular exercise and skin lubricants help in maintaining flexibility of limbs and pliability of skin.

Clinical use of colchicine :

- | | |
|--|--|
| 1. Acute gout, pseudogout. | 7. Psoriasis. |
| 2. Scleroderma. | 8. Behcet's syndrome, |
| 3. Primary biliary cirrhosis. | 9. Myelofibrosis. |
| 4. Amyloidosis. | 10. Erythema nodosum. |
| 5. Familial mediterranean fever. | 11. Sarcoid arthritis. |
| 6. Study of chromosomes (colchicine arrests cell division at the metaphase). | 12. Recurrent oral ulcers/pericarditis |
| | 13. Osteoarthritis. |

Case 63

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

Describe the face of the patient :

1. Classical photosensitive 'butterfly' rash over the malar areas and bridge of the nose (never involves the nasolabial folds)—slightly raised erythema, may be scaly.
2. The lesion consists of erythema and oedema during acute phase, atrophy and telangiectasia in chronic phase.
3. Patchy alopecia.
4. 'Lupus hairs'— Short, broken hairs seen above the forehead.
5. Ulcers within the oral cavity (palatal ulcer specially, usually painless and recurrent) and nasal mucous membrane (seen after opening the mouth or careful examination of the nose).

N.B. : The face with the 'butterfly' rash looks like the face of wolves ('Lupine' means, like a wolf). The patient is commonly a female one (F : M = 9 : 1) in her second or third decade of life. Butterfly rash in SLE is due to apoptosis of keratinized layer of skin exposed to sunlight/UV light.

* **Alopecia** is loss of hair in the scalp, and is of two types : Cicatricial (DLE) and non-cicatricial (alopecia areata, SLE).

Causes of erythematous butterfly-like lesion in face :

1. SLE.
2. Acne rosacea (due to vasodilatation and telangiectasia, there may be red patches with nose disfigurement or rhinophyma of nose; lesion increases after taking tea, coffee etc).
3. Discoid lupus erythematosus (DLE) or mixed connective tissue disease (MCTD).
4. Lupus vulgaris (commonest form of skin tuberculosis).
5. Photosensitive reaction (after ingestion of drugs like tetracycline or phenothiazines).
6. Chloasma or melasma.
7. Leprosy (lepromatous).
8. Dermatomyositis (heliotrope rash; specially the juvenile variety).
9. Post kala-azar dermal leishmaniasis (PKDL).
10. Pemphigus erythematosus (an uncommon variant of pemphigus).
11. Polymorphous light eruption.
12. Sarcoidosis.

Other skin lesions in SLE :

The skin is affected in 85% patients of SLE :

1. Ill-defined, erythematous macular or maculopapular rash with fine scaling (particularly in sun-exposed areas); photosensitivity.
2. Vasculitic lesions on the fingertips and around the nail folds, purpura.
3. Bullous lesion, hives; recurrent urticaria; lichen planus-like dermatitis.
4. Panniculitis (lupus profundus).
5. Periungual erythema.
6. Digital infarcts (watch the tips and pulps of fingers for any tiny cicatricial depression).
7. Livedo reticularis (cyanotic mottling of skin with fishnet appearance).
8. Raynaud's phenomenon.
9. Angioneurotic oedema (non-pitting oedema with itching).
10. Pigmentation.
11. Alopecia.

* Photosensitive rash occurs in face, ears, chin, anterior part of neck, upper back and extensor surfaces of arms.

** D/D of livedo reticularis in skin is erythema ab igne (reticular pattern due to prolonged exposure to heat) and cutis marmorata (physiological reaction to cold).

What is chronic discoid lupus erythematosus (CDLE) ?

Lupus erythematosus (LE) has three clinical variants like.

- Systemic lupus erythematosus (SLE).

- Chronic discoid lupus erythematosus (CDLE).
- Subacute cutaneous lupus erythematosus (SCLE).

CDLE is a chronic skin ailment which is commoner than SLE. The skin lesions heal with scarring, and the other characteristic features are :

1. Photosensitivity and erythema,
2. Scaling with atrophy,
3. Telangiectasia,
4. Hypopigmentation and keratotic plugging.

Sites - Face, arms, neck, scalp, ears.

* Rarely, in 5% patients DLE may turn into SLE, while DLE occurs in approximately 20% of patients with SLE. DLE is associated with positive ANF (30%) and normal complement level.

Causes of telangiectasias :

These are small, dilated blood vessels visible on skin surface; they blanch on application of pressure given by a pinhead.

- (A) Systemic diseases : SLE, scleroderma, dermatomyositis, hereditary haemorrhagic telangiectasis, ataxia-telangiectasia, carcinoid syndrome, spider naevi in cirrhosis of liver.
- (B) Primary cutaneous disorder : acne rosacea, ionising radiation, actinically damaged skin.

Common clinical features of SLE :

SLE is a multisystem disease. The common features are,

1. Arthritis and arthralgia (commonest).
2. Fever (presents as PUO).
3. Skin eruptions.
4. Lymphadenopathy.
5. Renal involvement.
6. Anorexia, nausea and vomiting.
7. Myalgia.
8. Pleurisy.
9. CNS abnormalities.

* Splenomegaly occurs in 20-30% and hepatomegaly in 25% patients of SLE.

Neurological manifestations of SLE :

1. Convulsions.
2. Depression.
3. Organic psychosis.
4. Cranial nerve involvement.
5. Transverse myelitis.
6. Organic brain syndrome (psychosis, seizure)
7. Migraine.
8. Chorea.
9. Cerebellar ataxia.
10. Rarely, peripheral neuropathy.
11. Cerebrovascular accidents.
12. Aseptic meningitis.

* Fundoscopy may reveal white, hard exudate called 'cytoid body'.

Cardiopulmonary involvement in SLE :

1. Pericarditis, myocarditis, non-bacterial verrucous endocarditis (Libman-Sacks endocarditis may produce a murmur).
2. Heart block and arrhythmia may occur.
3. Ischaemic heart disease (due to premature narrowing of coronary arteries).
4. Recurrent pleurisy and pleural effusion (exudate).
5. Lupus pneumonitis (pulmonary infection is very common).
6. 'Shrinking lung syndrome' (X-ray shows elevation of diaphragm and liner scars from recurrent pulmonary infarction).
7. Fibrosing alveolitis.
8. Intrapulmonary haemorrhage (from vasculitis).

What is drug-induced SLE :

1. Drugs with positive ANF — INH, phenytoin, phenothiazines, a-methyldopa and levodopa.
2. Drugs associated with exacerbations of SLE— Sulphonamides, penicillin and oral contraceptives.
3. Drugs producing syndrome similar to SLE— Hydralazine and procainamide.

N.B. : Anti-histone antibody is positive in drug-induced SLE. Renal and neurological manifestations are uncommon. Complement levels are normal.

Renal manifestations of SLE :

It carries the worst prognosis. Renal biopsy is an important guide to prognosis in SLE.

1. Minimal proteinuria, haematuria, cellular casts.
2. Nephrotic syndrome (minimal change to crescentic glomerulonephritis).
3. Chronic renal failure.
4. Urinary tract infections.

Classification of lupus nephritis :

It is done by International Society of Nephrology and Renal Pathology Society (2004). Clinical renal involvement occurs in approximately 30% cases. There are 6 classes :

Class I : Minimal mesangial lupus nephritis (LN).

Class II : Mesangial proliferative LN.

Class III : focal LN.

Class IV : Diffuse LN.

Class V : Membranous LN.

Class VI : Advanced sclerotic LN.

Causes of pain abdomen in SLE :

- | | |
|---------------------------|-----------------------------------|
| 1. Peritonitis. | 5. Complication of corticosteroid |
| 2. Pancreatitis. | therapy (peptic ulcer). |
| 3. Perisplenitis. | 6. Gall bladder perforation from |
| 4. Mesenteric vasculitis. | necrotising arteritis. |

* Death in SLE usually results from infection, cardiac failure or renal failure.

How to establish the diagnosis of SLE ?

1. Blood examination reveals normocytic normochromic anaemia or immune haemolytic anaemia, leucopenia, lymphocytopenia, thrombocytopenia, clotting factor defect (due to antibodies formed against factor VII, IX and X). ESR tends to be high with active disease.
2. Urine analysis - Proteinuria (may be in nephrotic range), haematuria, RBC casts.
3. LE cell phenomenon may be positive (not done now-a-days).
4. Coombs' test and rheumatoid factor (30%) may be positive.
5. False positive serological test for syphilis; positive lupus anticoagulant (LA).
6. Blood urea and creatinine may be high in renal involvement.
7. Antinuclear factor (ANF) are present in 98% cases. *Antibodies against native double-stranded DNA (70%) is specific for SLE. Anti-Sm antibody (Smith; 25%) is highly specific for SLE.*
8. Hypocomplementaemia C_3 (i.e., $C_3 < 5.5$ mg/dl).
9. Skin biopsy taken from uninvolved sun-hidden part (shows deposits of immunoglobulins and complement components at epidermal and dermal junction - called lupus band test' (60%))
10. Renal biopsy.

* It is to be remembered that C-reactive protein (CRP) is normal in SLE unless and otherwise there is superadded infection, pleurisy or arthritis (high CRP). Anticardiolipin antibody should be done to diagnose secondary antiphospholipid antibody syndrome.

Autoantibody associations in different subsets of SLE :

Autoantibodies are immunoglobulins which bind to antigens within autologous (self) tissue. Antinuclear antibody (ANA) or factor (ANF) are antibodies produced against nuclear component of a cell.

1. Anti-ds DNA—Nephritis, serositis.
2. Anti-Sm—Specific for SLE.
3. Anti-Ro/La—Photosensitivity, neonatal lupus.
4. Antiphospholipid antibody/Lupus anticoagulant—Coagulopathy, abortion, thrombocytopenia.
5. Anti-U1-RNP (nuclear ribonucleoprotein)—Mixed connective tissue disease.
6. Anti-histone Drug-induced lupus (e.g., hydralazine or procainamide-induced).
7. Anti-ribosomal P antibody—CNS lupus (psychosis, depression)

8. C autoantibody—having prognostic significance in lupus nephritis.
- * Other autoantibodies in different connective tissue diseases are :
 - Anti-centromere—CREST syndrome.
 - Anti-SSA (anti-Ro)/SSB (anti-La)—Sjogren's syndrome.
 - Anti-topoisomerase I (anti-Scl-70)—diffuse cutaneous scleroderma.
 - ANCA :
 - c-ANCA—Wegener's granulomatosis.
 - p-ANCA—Systemic vasculitis.
 - Anti Jo-1—Polymyositis, dermatomyositis with lung involvement.

How will you manage this patient ?

There is no cure for SLE. The aim of treatment is to maintain remission and control acute 'flares'.

1. Rest.
2. NSAID - For arthritis, arthralgia, myalgia, pleurisy, fatigue, fever etc.
3. Hydroxychloroquine - 400 mg/ day (ophthalmological examination at least once a year should be done and the patient should avoid ultraviolet light) for treatment of skin lesions and arthritis.
4. Low dose corticosteroid in debilitating skin lesion. Steroid is also indicated (60 mg/day of prednisolone) in cardiopulmonary, CNS and haematological involvement. Gradually, the dose is tapered after two weeks and the patient is maintained on 10 mg prednisolone/day.
5. Renal lesions need special mention. Renal biopsy should be done prior to treatment. Focal lupus glomerulonephritis respond well to treatment with prednisolone (40-60 mg/day till symptoms disappear). However, diffuse and membranous lesions do not respond well and they need therapy with 1 gm of I.V methyl prednisolone for 3 consecutive days followed by maintenance daily or alternate-day prednisolone (also used in organ damage in SLE). The widely accepted 'pulse' regimen goes like this : cyclophosphamide 500 mg/m², I.V, monthly for 6 months -> then 500 mg/m², I.V, 3 monthly for 2 doses -> mycophenolate mofetil (1 g BD/day) or azathioprine (1.5-3 mg/kg of body weight/day) for 2 years. Short-term studies show that mycophenolate is superior to cyclophosphamide in induction as well as maintenance phase. Other immunosuppressive drugs used are methotrexate, tacrolimus, cyclosporin. Rituximab (anti-CD20) is used in refractory cases to reduce the level of antibodies.
6. Infection anywhere in the body is treated by proper antibiotic.
7. Skin rash may be treated by sunscreen cream, retinoids or dapsone.
8. Plasmapheresis may be tried.
9. SLE with pregnancy is treated with lowest dose of prednisolone for the shortest period.
10. Antiphospholipid syndrome needs low dose aspirin and anticoagulant therapy.

What is antiphospholipid antibody syndrome (APS) or Hughes syndrome ?

In APS there is persistently elevated levels of anti-phospholipid/cardioliipin antibody and/or positive test for lupus anticoagulant, and the presence of one of the following i.e., arterial or venous thrombosis, recurrent foetal loss and thrombocytopenia. APS is of two types : primary (no cause found) and secondary (with SLE, rheumatoid arthritis, systemic sclerosis etc.).

Clinical features are recurrent arterial or venous thromboembolism, recurrent foetal loss, thrombocytopenia, livedo reticularis, valvular heart disease, chorea/migraine/epilepsy or catastrophic occlusion of blood vessels. APS is being treated with anticoagulants (heparin), antiplatelets (aspirin), IVIG and immunosuppressants.

Tests which are helpful in following the clinical course of SLE :

- 1) Titre of anti-ds DNA
- 2) Serum C₃ level
- 3) Haematocrit
- 4) ESR
- 5) WBC count
- 6) Platelet count
- 7) Urine analysis
- 8) Serum creatinine.

Indications for use of chloroquine in internal medicine :

- | | |
|--------------------------|---|
| 1. Malaria. | 5. Sjogren's syndrome. |
| 2. Hepatic amoebiasis. | 6. MCTD. |
| 3. Rheumatoid arthritis. | 7. Lepa reaction (type I). |
| 4. SLE and DLE. | 8. Congenital erythropoietic porphyria. |

Case 64

ERYTHEMA NODOSUM

Characteristic lesion :

- 1 • Single or multiple non-ulcerating, bluish-red or erythematous nodular lesion, usually in children or young adults; F > M.
2. Generally present on the extensor surfaces; shins are the usual site; thigh, upper arm or forearm rarely.
3. Painful and tender; at times bilateral and symmetrical; non-itchy.
4. May be associated with mild constitutional symptoms like fever, chills, malaise, arthralgia; may be recurrent.
5. Lesion is generally self-limiting (2-6 weeks). They fade through bruising; usually heal without scarring.

Common causes of erythema nodosum :

1. Tuberculosis (specially primary) —so, a chest radiography is essential.
2. P-haemolytic streptococcal infection (commonly after streptococcal sore throat).
3. Sulphonamide therapy.
4. Leprosy (lepromatous).
5. Sarcoidosis.
6. Ulcerative colitis or Crohn's disease.
7. Other drugs like oral contraceptives, barbiturates, penicillins, iodides and bromides.
8. Histoplasmosis or coccidioidomycosis, Behcet's syndrome.
9. Brucellosis, psittacosis, tularaemia.
10. Rarely in rheumatic fever.
11. Bacterial gastroenteritis (Salmonella, Shigella, Yersinia).
12. Idiopathic.

Basic pathology of the lesion :

Actually, their appearance reflects patchy inflammation of subcutaneous fat (panniculitis) and small blood vessels (vasculitis). This is an example of type III (immune-complex) allergic reaction.

Foreign proteins and bacteria are slowly removed from the 'front of the legs' due to poor lymphatic drainage (as tibia splints the overlying tissue, the massage of the lymphatic channels is reduced)—this is why shins are the usual site.

Erythema nodosum is a septal panniculitis and is not of vasculitic in origin.

Differential diagnosis :

- | | |
|------------------------|---|
| 1. Resolving bruises. | 4. Erythema chronicum migrans (Lyme disease). |
| 2. Thrombophlebitis. | 5. Erythema multiforme. |
| 3. Erythema induratum. | 6. Cutaneous vasculitis or metastases. |

Different allergic reactions of primary pulmonary tuberculosis :

1. Erythema nodosum,
2. Phlyctenular keratoconjunctivitis,
3. Some cases of pleural effusion (not accepted by all), and
4. Sometimes, pneumonitis from exudative hypersensitivity lesion.

Treatment :

1. Stoppage of the offending drug.
2. Treatment of the primary disease.
3. Analgesics (NSAID).
4. Lotio calamine paint over the lesion.
5. Prednisolone (upto 30 mg/day), potassium iodide, dapsone (100 mg/day) or colchicine (0.6 mg BD) in stubborn cases.

What is erythema nodosum leprosum (ENL) ?

1. 50% of lepromatous leprosy and 25% of borderline lepromatous leprosy patients suffer from this type of skin reaction during the course of their illness.
2. This is a type II lepra reaction.
3. Acute crops of painful, tender nodules appear along with fever, lymphadenopathy, arthritis and iritis. The lesion may necrose and discharge sterile pus. It may appear in any part of the body.
4. Treatment Is done mainly by thalidomide, 100 mg 4 times dally. The dose is gradually reduced over weeks or months. Thalidomide is strictly contraindicated in females of child bearing age due to its teratogenicity. If thalidomide Is contraindicated, prednisolone may be used. NSAIDs (as analgesic-antipyretic) are given, If necessary. The dose of clofazimine is increased to 200-300 mg daily for few weeks during the reaction period. Iritis is managed by local Instillation of 1% atropine sulphate.

Lesions characteristically found over the shin :

- | | |
|-------------------------|--------------------------------------|
| 1. Erythema nodosum. | 3. Necrobiosis lipoidica diabetorum. |
| 2. Pretibial myxoedema. | 4. Lichen amyloidosis. |

Identification points :

Single or multiple bluish-red nodular lesion in anterior aspect of one or both legs, specially in younger patients.

Case 65**SUBCUTANEOUS NODULES****Differential diagnosis :**

- | | |
|-----------------------|----------------------------|
| 1. Lipoma, fibroma. | 7. Cysticercosis. |
| 2. Neurofibroma. | 8. Osier's node. |
| 3. Rheumatoid nodule. | 9. Tophi. |
| 4. Rheumatic nodule. | 10. Xanthoma. |
| 5. Leprosy. | * 11. Metastatic carcinoma |
| 6. Calcinosis. | 12. Panniculitis. |
- * Skin deposits from carcinoma, lymphoma and leukaemias.

Common sites for rheumatoid nodule :

These nodules signify underlying vasculitis and

1. Most commonly over the extensor surface of elbows.
2. Back of the head.
3. Over the sacrum in bed-ridden patients.

active disease. They are situated :

4. Extensor tendons of fingers and toes
5. Over the scapula.
6. Achilles tendon.
7. Margins of patella.

Common sites for rheumatic nodule :

The distribution is more or less like rheumatoid nodules i.e., situated over the bony prominences or at the site of pressure points while lying in bed.

Table 29 : Differentiation between rheumatoid and rheumatic nodules

Features	Rheumatoid	Rheumatic
1. Size	Big	Relatively smaller
2. Ulceration	Frequently occur	Does not ulcerate
3. Secondary Infection	May occur	Not common
4. Tenderness	Non-tender	May be tender
5. Fixity to skin	Very often fixed	Skin Is free
6. Incidence	20%	3-5% In India
7. Association	Indicates active disease with +ve RF	Associated with active carditis

Case 66

DOWN'S SYNDROME (MONGOLISM)

What is Down's syndrome ?

It is the commonest chromosomal disorder (1 in 700 newborns). Chromosome 21 is present in triplicate (i.e., total 47 chromosomes) as a result of non-disjunction during meiosis and is known as 'trisomy-21'.

Describe the facies (mongol facies) :

1. Small, round and flat face, brachycephaly; with downy forehead and short, wide fleshy neck.
2. Upwards-slanting eyes (oblique orbital fissure) with epicanthic folds at inner angles.
3. Low set ears; ears are small and dysplastic.
4. Small nose with depressed bridge of the nose.
5. Hypertelorism (widely set eyes).
6. High arched palate with small teeth.
7. Open mouth with protruded and furrowed tongue (macroglossia), with
8. An idiotic look (cheerful idiot).

* There are Brushfield's spot on the iris and typical facial grimace on crying.

** It is often said that the tongue is normal but it protrudes because of small size of the oral cavity.

*** John Langdon Hayden Down (Langdon-Down) was Physician (1828-1896), The London Hospital, London who first described the syndrome in 1866.

**** Epicanthic folds may be seen in Turner's syndrome.

Diagnostic points in hand, foot and eye ;

(A) Hand—

1. Short and broad hands (simian hand); short, stubby spatula-like fingers.
2. Single palmar flexion crease (simian crease).
3. Clinodactyly (hypoplasia of middle phalanx of 5th finger produces 'incurving' little finger).
4. Missing of one crease in little finger (cause—same as point 3).
5. Characteristic dermal markings in fingers (dermatoglyphics)—More than eight ulnar loops on fingers, and the ATD angle (distal axial triradii) becomes obtuse.

(B) Foot—

1. Increased gap between first and second toe (sandal gap).
2. Sub-hallucial pad of fat.
3. Single, deep and longitudinal crease in the sole of feet.

(C) Eye—

1. Brushfield's spot (whitish speckling on the iris).
2. Sometimes, cataract is present.
3. Rarely, squint and high myopia may be observed.

Features in other systems in Down's syndrome :

- (A) CVS— Endocardial cushion defects, e.g., VSD, ASD, PDA.
- (B) Gastrointestinal system— Duodenal stenosis, biliary atresia, severe periodontal disease.
- (C) Haematopoietic— There is many fold higher incidence of acute leukaemia (AML in the new-born and ALL in older children).
- (D) Neuromuscular— Hypotonia (poor Moro's reflex), floppiness in neonate.
- (E) Skeletal— Short-height, joint hyperextensibility.
- (F) Personality— Very pleasant with clean habits, and **fond of music**.
- (G) Intelligence— I.Q. is generally low (from mild to severe) and often very much mentally retarded. Speech and social developments are severely affected.

* Patients of Down's syndrome may develop early Alzheimer's disease in the 3rd or 4th decade.

** Coeliac disease may be associated with Down s syndrome.

Cause of death :

1. Recurrent respiratory tract infections,
2. Septal defects in heart, or
3. Acute leukaemia (usually AML).

Management :

There is no definite treatment for Down's syndrome. Management is aimed at treatment of respiratory tract infections, acute leukaemia, VSD or proper rehabilitation. They are usually trainable children but may be educable rarely.

* Down's syndrome and Klinefelter's syndrome are associated with increased maternal age (> 35 years) whereas, Marfan's syndrome and achondroplasia are seen with increased paternal age. Prenatal diagnosis of Down's syndrome may be done by amniocentesis or chorionic villus biopsy.

Identification points :

Typical facies, grimace and behaviour.

Case 67**HYDROCEPHALUS****What is hydrocephalus ?**

This is the dilatation of ventricles of brain due to increase in the volume of CSF within the ventricles resulting from a disturbance in the circulation (of CSF). It may be due to obstruction of CSF circulation or there may be failure of CSF absorption by the arachnoid villi.

Diagnosis of a child with hydrocephalus :

1. Abnormal enlargement of head, specially the frontal area.
2. Sutural separation.
3. Bulging of the anterior fontanelle.
4. Setting sun' sign— The eyes appear to be pushed down and thus both the upper bulbar conjunctiva become visible.
5. Scalp veins are dilated, and the skin over the scalp is thin and shiny.
6. Macewen sign— Skull is resonant (cracked-pot) on percussion.
7. High pitched cry.

Circulation of CSF :

Choroid plexus of lateral ventricles (major source of CSF formation)—Foramen of Monro—Third ventricle—Aqueduct of Sylvius—Fourth ventricle in the medulla—Foramen of Magendie and Luschka—Cisterna magna and cisterna pontis—Subarachnoid space. CSF is absorbed by arachnoid villi.

Types of hydrocephalus :

(A) Compensatory : Due to cerebral atrophy from any cause.

(B) Hypertensive :

- a) Obstructive If the circulation of CSF is obstructed within the ventricular system.
- b) Non-obstructive or communicating (i.e., obstruction distal to fourth ventricle)—Hydrocephalus resulting from occlusion of the subarachnoid cisterns (due to arachnoiditis) or interference with the CSF absorption.

Features of late onset hydrocephalus :

1. Head size may not enlarge.
2. Sutural separation may not be evident.
3. Mental retardation.
4. Presence of papilloedema.
5. Spastic and ataxic child (legs are more involved).
6. Urinary incontinence.

Aetiology of hydrocephalus :

1. Meningitis (bacterial or tuberculous).
 2. Head injury.
 3. Subarachnoid haemorrhage.
 4. Sagittal sinus thrombosis.
 5. Brain tumours (posterior fossa).
 6. Cerebral haemorrhage.
 7. Congenital e.g.. Arnold-Chiari malformation.
 8. 'Normal pressure hydrocephalus.
 - 9- Cerebellar abscess or haemorrhage.
 10. 3rd ventricle colloid cyst.
- * Obstructive 5, 6, 7, 9 and 10: non-obstructive—1, 2, 3, 4 and 8; 1, 2 and 3 are responsible for majority of cases.

Causes and features of acute hydrocephalus :

Causes are :

1. Head injury with its complications.
2. Subarachnoid haemorrhage.
3. Acute exudative meningitis.

Features are :

1. Severe headache and vomiting.
2. Drowsiness, convulsions or coma.
3. Papilloedema may be evident.
4. 6th cranial nerve palsy (false localising sign).
5. Jerks— Brisk (may be lost).
6. Plantar response Extensor.

What is 'normal pressure' hydrocephalus ?

It is a misnomer. Initially there is increased pressure but ultimately CSF haemodynamics compromise (ventricles dilate) and it becomes normal or low pressure hydrocephalus. Actually, it is a variety of communicating hydrocephalus. This rare condition is predominantly seen in old age with the triad of symptoms such as **dementia, ataxia and urinary incontinence**. Though it may result from head injury, subarachnoid haemorrhage or meningitis, many a time no cause is identified.

How to diagnose hydrocephalus very early ?

By serial recording of head circumference of the child. An increase in the head circumference by more than 1 cm in every 2 weeks for the first 3 months of life, makes the pediatrician suspicious.

What is normal intracranial pressure (ICP) ?:

It is 5-10 mm of Hg when measured indirectly. Now-a-days, devices measure ICP directly by implanting into the lateral ventricles. When ICP is > 15 mm of Hg, it is known as raised intracranial pressure.

What is benign intracranial hypertension (BIH) ?

Previously the term BIH was used synonymously with pseudotumour cerebri, meningeal hydrops or otitic hydrocephalus. It is a benign or idiopathic raised ICP developed as a result of diffuse swelling of brain with dilated ventricular system. The patient characteristically presents with headache, vomiting and papilloedema. BIH commonly develops from corticosteroid withdrawal, tetracycline or nalidixic acid therapy, hypervitaminosis A, Addison's disease, hypoparathyroidism, or COPD with type II respiratory failure. The CSF is under increased pressure but its analysis remains normal. CT scan is within normal limit except the dilated ventricles. Acetazolamide or repeated lumbar puncture may be of some help.

Diagnosis of hydrocephalus by investigations :

1. X-ray of skull-
 - (i) Enlargement of calvarium.
 - (ii) Sutural diastasis (separation).
 - (iii) Thinning of bone.
 - (iv) Erosion of clinoid process.
 - (v) Deepened sella turcica.
2. Ventriculography or cisternography by a suitable contrast medium (to see the site of obstruction).
3. Pneumoencephalography may be tried to localise the site of obstruction not done now-a-days.
4. CT or MRI scan (to show enlarged ventricles and cortical atrophy).
5. CSF flow studies may be done (CSF analysis is not informative).
6. Measurement of ICP—diagnostic.

Differential diagnosis of hydrocephalus :

1. Megalo encephaly (with severe mental retardation, and absence of IIT).
2. Chronic subdural haematoma (mostly with enlargement of the parietal regions).

Treatment of hydrocephalus :

It is important to treat the child before irreparable damage sets in.

(A) Drug—Acetazolamide (50-75 mg/kg/day) for reduction of CSF production.

(B) Shunt surgery—

- (i) Ventriculo-peritoneal (commonly used).
- (ii) Ventriculo-atrial.
- (iii) Ventriculo-caval (to jugular vein or superior vena cava).
- (iv) Lateral ventricle to cisterna magna.
- (v) Ventriculo-ureteric.

Different varieties of silastic valves are used and the commonly used one is 'Spitz-Holter' valve.

(C) Miscellaneous Compressive head wrapping, repeated lumbar puncture (in acute hydrocephalus), different diuretics, steroid, isosorbide are tried. Neurosurgical removal of tumour, if necessary.

Complications of 'shunt' :

1. Shunt closure— Displacement, kinking or plugging of the shunt.
2. Ventriculitis (by *Staphylococcus albus*).
3. Bacterial colonisation of the shunt.
4. Shunt nephritis (acute glomerulonephritis).
5. SVC obstruction.

How the head circumference of a child is measured ?

Ideally by a steel tape. The tape is passed over the most prominent part of the occiput and the forehead above the supraorbital ridges.

* Always measure the head circumference in a child with apparent enlargement of head.

Case 68**RICKETS****Clinical manifestations :**

(A) Symptoms :

1. Fretful, irritable, pale child with flabby muscles in the age group of 4 months to 2 years.
2. Delayed development; often late eruption of teeth.
3. Failure to sit, crawl, stand and walk at the normal ages.
4. Recurrent respiratory and G. I. tract infections.
5. Rocking of head in pillows with sweating in forehead (during sleep).
6. Poor dentition with late eruption of teeth.
7. Manifestations of tetany may develop.

(B) Signs :

a) Head :

1. *Craniotabes (thin and deformed skull)*—Earliest manifestation of rickets-, when the occipital or parietal bone is compressed and released, there is feeling like compressing a ping pong ball (egg-shell crackling feeling). It is seen below the age of 1 year and is due to small islands of unossified area in the skull.
2. Frontal and parietal bossing with large head (philosopher's head).
3. Quadrate skull with 'hot-cross bun' appearance; skull is apparently larger than normal.
4. Widening of sutures.
5. Late appearance of temporary teeth.
6. Delayed closure of anterior fontanelle.

* Other causes of *craniotabes* are hydrocephalus, congenital syphilis and osteogenesis imperfecta.

b) Chest:

1. Pigeon chest (pectus carinatum).
2. Rickety rosary (swelling or beaded appearance of the costochondral junction).
3. Harrison's sulcus (linear transverse depression by the side of xiphoid process).
4. Kyphoscoliosis.

c) Abdomen :

Pot-belly— Due to multiple factors like weakness of abdominal muscles, gaseous distension, hepatosplenomegaly, visceroptosis or lumbar lordosis.

d) Extremities :

1. Epiphyseal enlargement at wrists and ankles.
2. 'Knock knee' or 'bow leg' (when the child starts walking and thus, unusual before 1 year of age).
3. Genu valgum, varum or recurvatum (acrobatic rickets).
4. Pelvic deformities (triradiate pelvis—occurs in later childhood and may produce difficulties during childbirth).

* Rickets develops as a result of vitamin D deficiency in growing children before the fusion of epiphyses.

How to diagnose rickets ?

(A) Classical clinical features.

(B) Radiological changes (of upper extremity) :

1. Cupping, widening and fraying (saucer deformity) of lower end of radius and ulna.
2. Diminution of bone density.
3. Widening of wrist (soft tissue shadow).
4. Increased distance between the distal end of bones of forearm and metacarpal bones because of non-calcification of the rachitic metaphysis.

(C) Biochemical changes :

1. Plasma alkaline phosphatase is raised (of diagnostic value)—indicates osteoblastic activity.
2. Low phosphorus (due to associated secondary hyperparathyroidism).
3. Plasma calcium tends to fall from its normal value (usually remains normal).
4. Plasma 25-hydroxyvitamin D₃ level is low in majority of patients.

* Clinical rickets may occur with normal level of plasma calcium and phosphorus.

** Normal serum alkaline phosphatase level : a) 2-4.5 Bodansky units, b) 4-13 King-Armstrong units, c) 50-130 International units/per liter.

*** Remember, in health, alkaline phosphatase is elevated during skeletal growth.

What are the early manifestations of rickets ?

- | | |
|---|-----------------------------|
| 1. Irritability and restlessness. | 4. Rickety rosary. |
| 2. Head rolling and sweating in forehead, specially during sleep. | 5. Big anterior fontanelle. |
| 3. Craniotabes. | 6. Swelling of wrists. |
| | 7. Delayed dentition. |

Radiological evidences of 'healing rickets' :

1. The density of metaphysis is increased and roughness is diminished.
2. Epiphyseal cartilage becomes horizontal.
3. Horizontal 'Harrison's line' are seen in the lower end of diaphysis (initially near the epiphysis).

Management of rickets :

1. Vitamin D — 1000-2000 I.U. orally daily, depending on the age of the child and severity of the disease, for 6 to 12 weeks followed by daily supplement of 400 I.U. of vitamin D (**standard daily requirement of vitamin D is 400 I.U or 10 pig**). Few clinicians prefer larger doses; 500-1000 mg calcium is prescribed daily. Therapeutic dose is continued so long the alkaline phosphatase level remains high (normalisation of alkaline phosphatase is a good indicator of healing).
2. Diet— Must be rich in vitamin D and calcium e.g., milk, cod liver oil, halibut liver oil, fatty fish, egg, liver, fortified margarine should be taken.
3. Calcium may be given in the form of lactate or gluconate.
4. Treatment of **tetany** is done by inj. calcium gluconate (10% solution) 20 ml in normal saline

by I.V route, slowly to run over 4-8 hours (inj. calcium chloride may be given). If tetany is not controlled by calcium, administration of I.V magnesium may be necessary. Correction of alkalosis should also be done.

5. If after administration of vitamin D, no sign of radiological healing is seen within 4 weeks, the dose is increased to several thousand I.U. If no healing is observed even after this larger dose, 'refractory rickets' should be considered.
6. The child should be encouraged to play outdoors for exposure to sunlight.

What is 'refractory rickets'?

Definition : These patients develop rickets inspite of receiving the preventive daily dose (400 I.U) of vitamin D and do not show signs of healing rickets in X-ray, obtained 2 weeks after the parenteral administration (I.M) of 2 doses of 6 lakhs I.U. of vitamin D given at 15 days interval. The causes are :

1. Renal :
 - (i) Renal tubular acidosis.
 - (ii) Fanconi's syndrome.
 - (iii) Familial hypophosphataemic rickets (X-linked dominant disease with renal tubular loss of phosphate).
2. Malabsorption (e.g., coeliac disease).
3. Hepatic dysfunction— Impairs 25-hydroxylation of vitamin D.
4. Miscellaneous—
 - (i) Hypophosphatasia (deficiency of alkaline phosphatase).
 - (ii) Children on long term antiepileptic drugs.
 - (iii) Certain tumours may secrete parathormone like substances.

* Alkaline phosphatase is low in one variety of rickets only— Rickets due to hypophosphatasia.

** For 'causes of rickets', read 'Bedside Clinics in Medicine, Part II'.

Treatment of refractory rickets :

(A) Large dose as much as 15 lakhs I.U. of vitamin D, I.M is administered daily and subsequently, 3 lakhs I.U. of vitamin D is used daily as maintenance dose. In the presence of renal disease, i.ring of 1 u-hydroxycholecalciferol may be given daily, orally.

- (B) Administration of neutral phosphate salts, and
- (C) The treatment of the primary cause.

What is vitamin D₂ and D₃ ?

Vitamin D₂ is ergocalciferol, and vitamin D₃ is cholecalciferol.

Commonest cause of vitamin D deficiency in India :

Malabsorption.

What is hypervitaminosis D ?

Chronic oral intake of large dosage of vitamin D (50000-100000 I.U daily) for months may give rise to hypotonia, anorexia, nausea, vomiting, Irritability, constipation, abdominal pain, failure to thrive and pallor. Hypervitaminosis D is associated with high serum calcium level. Hypercalcaemia ultimately leads to renal damage and eventually metastatic calcification develops.

Treatment :

1. Administration of vitamin D is stopped immediately.
2. EDTA, sodium phytate may be used.

Megavitamin question—vitamins responsible :

There may be 'hypervitaminosis' with :

1. Vitamin A— Anorexia, nausea, vomiting, pain abdomen, bone pain, weight loss, cracked lips and benign intracranial hypertension, hyperostosis, hair loss and desquamation of skin. High doses of carotenoids are avoided in smokers due to an increased risk of lung cancer, and in hypothyroid patients who may develop carotenaemia.
2. Vitamin D— Mentioned above.
3. Vitamin C— Oxalate stone, scurvy in the offspring, increased intestinal absorption of iron.
4. Pyridoxine or vitamin B₆— Sensory polyneuropathy.

5. Vitamin K— Jaundice in newborn if given to pregnant women.
6. Vitamin E— Malaise, headache, G. I. ailments and reduced platelet aggregation.
7. Niacin—Flushing, pruritus, aggravation of asthma, gout and acanthosis nigricans.

When the fontanelles close normally ?

Anterior fontanelle closes by 9-18 months. Posterior fontanelle closes by 3-6 months.

Delayed closure of anterior fontanelle :

- | | |
|-------------------------|---------------------------|
| 1. Malnutrition. | 5. Hydrocephalus. |
| 2. Rickets. | 6. Down's syndrome. |
| 3. Congenital syphilis. | 7. Mucopolysaccharidosis. |
| 4. Cretinism. | |

Bulged anterior fontanelle :

1. Crying infant.
2. Increased intracranial tension (e.g., meningitis, brain tumour).
3. Hydrocephalus.
4. Benign intracranial hypertension (corticosteroid withdrawal, vitamin A overdose, tetracycline therapy, hypoparathyroidism etc).
5. Galactosaemia.

High arched palate :

- | | |
|-----------------------|---------------------------------------|
| 1. Down's syndrome. | 3. Cyanotic congenital heart disease. |
| 2. Marfan's syndrome. | 4. Turner's syndrome. |

* Keep the patient's mouth wide open and look for palate keeping the eyes at the level of patient's upper incisor teeth—if the roof of palate is not visible, it is known as high arched palate.

Frontal bossing (prominence) of skull :

- | | |
|-------------------------|----------------------------|
| 1. Thalassaemia major. | 5. Achondroplasia. |
| 2. Rickets. | 6. Acromegaly. |
| 3. Congenital syphilis. | 7. Ectodermal dysplasia. |
| 4. Hydrocephalus. | 8. Ehlers-Danlos syndrome. |

Depressed bridge of the nose :

- | | |
|-------------------------|--|
| 1. Racial. | 7. Wegener's granulomatosis. |
| 2. Cretinism. | 8. Ectodermal dysplasia. |
| 3. Thalassaemia major. | 9. Achondroplasia. |
| 4. Down's syndrome. | 10. Osteopetrosis (marble bone disease). |
| 5. Lepromatous leprosy. | 11. Hurler syndrome. |
| 6. Congenital syphilis. | 12. Midline granuloma. |

Upwards-slanting (mongoloid) of eyes :

- | | |
|---------------------|---------------------------|
| 1. Racial. | 3. Ectodermal dysplasia. |
| 2. Down's syndrome. | 4. Prader-Willi syndrome. |

Anti-mongoloid slant of eyes :

- | | |
|-----------------------|-----------------------|
| 1. Noonan's syndrome. | 3. Turner's syndrome. |
| 2. Apert's syndrome. | 4. Alport syndrome. |

Hypertelorism (widely set eyes) :

- | | |
|------------------------|---|
| 1. Racial. | 5. Craniostenosis (e.g., Crouzon syndrome). |
| 2. Down's syndrome. | 6. Turner's syndrome. |
| 3. Cretinism. | 7. Ehlers-Danlos syndrome. |
| 4. Thalassaemia major. | 8. Noonan's syndrome. |

* Diagnosed clinically by measuring inter inner-canthal distance between two eyes which is more than half of the inter-pupillary distance, or inter-pupillary distance > 85 mm where the average normal value is 60 mm. Hyperplasia of lesser wing of sphenoid bone is usually responsible for hypertelorism.

Hypotelorism :

It means there is decreased distance between orbits, and is found in ethmocephaly, cebocephalia or Patau's syndrome.

Sabre tibia (anterior tibial bowing) :

- | | |
|--|-----------------------------|
| 1. Congenital syphilis. | 3. Rickets. |
| 2. Paget's disease (osteitis deformans). | 4. Osteogenesis imperfecta. |
| | 5. Osteopetrosis. |

* 'Sabre' literally means cavalry sword with curved blade.

Prominent supraorbital ridges :

- | | |
|---------------------|--------------------|
| 1. Racial. | 4. Achondroplasia. |
| 2. Acromegaly. | 5. Hydrocephalus. |
| 3. Paget's disease. | 6. Thalassaemia. |

Low set ears :

- | | |
|---------------------|------------------------------------|
| 1. Down's syndrome. | 3. Trisomy-13 (Patau's syndrome). |
| 2. Elfin facies. | 4. Trisomy-18 (Edward's syndrome). |

* Normally, $\frac{2}{3}$ rd of the length of pinna is seen above an imaginary horizontal line, extending from outer canthus of eye to ipsilateral pinna. If less than $\frac{2}{3}$ rd of length of pinna is seen above that line, low set ears are said to be present.

Deep ear lobe cleft :

Though occasionally congenital, the prominent diagonal crease seen over the ear lobe is now well-recognised as strong and independent risk factor for coronary heart disease.

Quadrate skull :

1. Rickets.
2. Osteopetrosis.

Short neck :

- | | |
|--------------------------------|-----------------------|
| 1. Cranio-vertebral anomalies. | 4. Turner's syndrome. |
| 2. Cretinism. | 5. Hurler syndrome. |
| 3. Down's syndrome. | 6. Noonan's syndrome. |

Identification points (rickets) :

- | | |
|--|-------------------------------|
| 1. Fretful child. | 4. Hypotonia. |
| 2. Apparently large head. | 5. Pot-bellied abdomen. |
| 3. Swelling of lower end of wrists and ankles. | 6. 'Knock knee' or 'bow leg'. |

Case 69**MYXOEDEMA****What is myxoedema ?**

Myxoedema results from failure of the thyroid gland (primary disease of the thyroid) after puberty and is characterised by wide-spread deposition of mucinous material (mucopolysaccharides, hyaluronic acid and chondroitin sulphate) in different parts of the body, and clinically manifested by poverty of physical, mental and metabolic activities.

Describe the facies in myxoedema (hypothyroidism) :

1. Dull expressionless, puffy face (periorbital puffiness with baggy lower eyelids); xanthelasma.
2. Coarse hair; fall of hair in scalp (patchy alopecia) and lateral 1/3rd of eyebrows (madarosis).
3. Dry, rough and coarse skin.
4. Facial pallor (sometimes, rose-purple malar flush is seen); thick lips and macroglossia may be present.
5. Hoarseness of voice with bradylalia.

* Don't forget to look at the swollen neck and fullness of the supraclavicular fossa during inspection of face. It is one of the D/D of moon *face as well as masked face*.

** 'Bags under the eyes' are often seen in insomnia, senility, chronic alcoholism, prolonged crying and myxoedema. It may not have any clinical significance.

*** *Madarosis* is common in leprosy, myxoedema, amyloidosis and neurodermatitis.

Is there any goitre ?

Usually thyroid tissue is not readily palpable in myxoedema. If palpable, think of :

1. Hashimoto's thyroiditis,
2. Iodine deficiency,
3. Use of antithyroid drugs, and
4. Malignancy of the thyroid gland.

Why do you say myxoedema ?

Myxoedema is common in females and there are presence of the following :

(A) Common symptoms are (if you are allowed to take the history) :

- | | |
|--|--|
| (i) Tiredness, lethargy, somnolence; hoarse voice. | (vi) Effort angina. ' |
| (ii) Weight gain; non-pitting swelling of legs. | (vii) Loss of libido. |
| (iii) Cold intolerance. | (viii) Stiff and aching muscles; arthralgia. |
| (iv) Constipation. | (ix) Deafness. |
| (v) Menorrhagia or irregular bleeding. | (x) Repeated abortions in productive age. |

(B) The signs are :

- (i) Physical sluggishness.
- (ii) Dry, cold, rough or coarse skin with lustreless, brittle and scanty hairs. There may be lemon-yellow tint of the skin due to carotenaemia.
- (iii) Moderate to severe anaemia.
- (iv) Deep and *hoarse voice* (due to myxomatous infiltration of vocal cord); bradylalia; macroglossia.
- (v) Sinus bradycardia.
- (vi) Obesity with non-pitting oedema or solid oedema of legs (supraclavicular fossa, neck and limbs are swollen more).
- (vii) Delayed relaxation of ankle jerk (it is better to demonstrate by 'special method' where the patient has to kneel-down on a chair).

* One must examine all the points.

** *Bradylalia* is also seen in depression and parkinsonism.

*** *Heat intolerance* is seen in hyperthyroidism and menopause.

**** Other common causes of *hoarse voice* are laryngitis, singer's nodule, voice-abuse, excessive use of tobacco, recurrent laryngeal nerve palsy, bulbar palsy.

Why there is anaemia ?

1. Lack of thyroxine— Normocytic normochromic type.
2. Menorrhagia— Iron deficiency anaemia.
3. Sometimes, myxoedema is associated with pernicious anaemia.

Mention causes of 'pallor without anaemia' :

1. Peripheral circulatory failure (e.g., low cardiac output in acute LVF) or shock (ashen-grey pallor), or vasoconstriction due to any cause; syncope.
2. Acute myocardial infarction.
3. Very tight aortic stenosis or mitral stenosis.
4. Myxoedema (pallor > anaemia)— Pallor is due to vasoconstriction plus lemon-yellow tint due to carotenaemia, plus anaemia.
5. Nephrotic syndrome.
6. Sheehan's syndrome or panhypopituitarism.
7. Vasovagal attack, fear, exposure to cold, intense emotion; increased carotenoid pigment.
8. Night workers.
9. Oedematous conditions, e.g., anasarca.
10. Thick skin (e.g., scleroderma).

- * **Pallor** may be associated with or without anaemia :
- Pallor with anaemia— All types of severe anaemia, SBE etc.
 - Pallor without anaemia— Described above.

Mention causes of constipation in clinical practice :

(A) Acute : Intestinal obstruction; acute abdomen.

(B) Chronic :

- Faulty habits, lack of physical activity, less intake of fibre; less access to toilet facility.
 - Painful anal conditions e.g., anal fissure; proctitis.
 - Obstructive lesions like carcinoma, stricture.
 - Disturbed motility : irritable bowel syndrome, systemic sclerosis, pregnancy, myopathy, Chagas' disease, parkinsonism, spinal injury.
 - Metabolic : Hypothyroidism, hypokalaemia, hypercalcaemia.
 - Drugs : Antidepressants, codeine, iron and calcium supplements, aluminium containing ant-acid, calcium channel blockers.
- * Though difficult to define, constipation is the infrequent passage of hard stools or stool frequency < 3 times/week (highly subjective).

Cardiovascular signs in myxoedema :

- Pulse— Sinus bradycardia.
 - BP— Systolic pressure may be high due to hypercholesterolaemia and atherosclerosis (with or without xanthelasma). There may be diastolic hypertension.
 - Features of CCF (rare).
 - Cardiomyopathy.
 - Features of pericardial effusion, rarely.
- * Patient may complain of anginal pain or exertional shortness of breath.

Why there is cardiomegaly in myxoedema ?

- Cardiac dilatation, 2) Pericardial effusion, or 3) Cardiomyopathy.

Dermatological manifestations in myxoedema :

Non-pitting oedema of hands and feet, periorbital puffiness, dry and lustreless skin with alopecia, purplish lips, malar flush, xanthelasma, carotenaemia and occasionally erythema ab igne.

Features in nervous system :

- Higher function—
 - Poor memory, delayed cerebration.
 - Psychosis (myxoedema madness).
 - Depression.
 - Myxoedema coma.
 - Mental confusion, convulsions due to syndrome of inappropriate ADH secretion (SIADH) - Rare.
- Speech— Slow (bradylalia), sluggish and hoarse.
- Cranial nerves— Nerve type of deafness.
- Cerebellar ataxia.
- Hypotonia.
- Ankle Jerk— Delayed relaxation** (myotonic reflex); at times there is 'hung-up reflex'.
- Entrapment neuropathy— Carpal tunnel syndrome.
- Myotonia (Hoffman's syndrome)— Painful cramps in muscles; proximal myopathy
- Muscle stiffness, and aches and pains in muscle; percussion myoedema (a small ridge of temporary swelling at the point of stimulation by hammer).
- Rarely, pseudomyotonia.
- Peripheral neuropathy rarely.
- Unmasking of myasthenia gravis.
- Hypokalaemic periodic paralysis.



A patient of **chorea** at the onset of choreiform movement



Tetanus with a fixed sardonic smile (risus sardonicus or smile of 'Satan') – the smile does not reach the eyes



"Main d' accoucheur" or carpal spasm in **tetany**



Catatonic patient (hypertonia) holding a rigid posture for hours. It is the admixture of psychic and motor disturbance



Multiple neurofibroma



Gibbus – acute angulation of vertebra; here it is due to metastasis from lung



Pectus excavatum or funnel chest



Emaciation evidenced by wasting (eg, supraspinatus), winging of the scapulae (prominent medial border) and vertebral prominence in pulmonary tuberculosis



Pectus carinatum or pigeon chest. Rickety rosary and Harrison's sulcus are seen in a patient of rickets



Kyphoscoliosis with winging of the scapula

What is myxoedema coma ?

It is a rare but dangerous presentation of myxoedema, seen usually in elderly during winter months, due to acute thyroid hormone deficiency and is characterised by,

1. Hypothermia (body is cold and dry without any shivering)—temperature as low as 80°F,
2. Hypotension,
3. Hyponatraemia,
4. Hypoventilation,
5. Hypoxia,
6. Hypoglycaemia,
- 7- Bradycardia,
- 8- Convulsions,
- 9- Unconsciousness.

There is very high mortality (50%) and survival depends upon early recognition and treatment.

What is subclinical hypothyroidism?

The patient is asymptomatic and clinically euthyroid but biochemically T_3 and T_4 are in the lower limit of normal with high TSH.

Carotenaemia (carotenoderma) and its clinical associations .

Carotenaemia is the presence of increased amount of carotene within the body. It is seen in •.

1. Eating large quantities of carrots, oranges, papaya, mango and other vegetables e.g., squash.
2. Hypothyroid (due to impaired metabolism of carotene in the liver).
3. Diabetes mellitus.
4. Receiving p-carotene for the treatment of erythropoietic porphyria.
5. Anorexia nervosa.
6. Simmond's disease.
7. Castrated male.

It produces an orange-yellow or lemon-yellow colour of skin specially of face, palms and so es. Sclera never turns yellow (D/D of jaundice).

Table 30 : Differentiation between primary and secondary hypothyroidism :

Features	Primary	Secondary
1. Skin	Thick and without wrinkles	Thin with fine wrinkles
2. Hair	Coarse	Fine
3. Menstrual irregularities	Menorrhagia	Amenorrhoea
4. Secondary sexual characters	Normal	Poor
5. Heart size	May be enlarged	Small
6. Goitre	May be present	Absent
7. Soft tissue oedema	Marked	Absent
8. Blood pressure	Normal or high	Low
9. Cholesterol	Increased	Normal
10. TSH	High	Low
11. Plasma cortisol	Normal	Low
12. TRH stimulation test	Exaggerated response	No response
13. Thyroid autoantibodies	May be present	Absent

* TO DIAGNOSE PRIMARY AND SECONDARY HYPOTHYROIDISM CLINICALLY, ONE SHOULD EXAMINE THE SKIN, HAIRS, SECONDARY SEXUAL CHARACTERS AND SOFT TISSUE OEDEMA.

What is juvenile myxoedema ?

In this condition, hypothyroidism starts during childhood and adolescence period (before puberty) but the thyroid function is normal at birth or infancy. There is delayed puberty, growth retardation with delayed bone age and galactorrhoea.

How will you investigate the case ?

1. Blood—
 - a) Low Hb (usually normocytic normochromic, or macrocytic) and low RBC count.
 - b) Cholesterol— High (> 200 mg/dl).
2. Chest X-ray— Cardiac shadow may be enlarged
- 2 ecg—
 - a) Low voltage (R-waves in limb leads are < 5 mm and that in precordial leads are <10 mm).

- b) Sinus bradycardia.
- c) Non-specific ST-T changes.
4. Plasma TSH estimation—Elevated in myxoedema (normal value 0-3-5 nU/ml).
5. Plasma T₃ and T₄ estimation—Low in myxoedema.
(normal value of T₃ is 80-150 ng/100 ml and that of T₄ is 4-12 jig/100 ml).
6. Radioactive Iodine 131 uptake by the thyroid gland is reduced.

now-a-days, ram (InStmmntal ^cording of jerks)- Delayed relaxation of ankle jerk; not used

8. Muscle enzymes—Elevated LDH, SGOT (AST) and CK.
9. FNAC and USG of thyroid gland in selective cases may be helpful.

* **T₄ or T₃ Peroxidase (TPO) antibodies** may be demonstrated in Hashimoto's thyroiditis. **T₄ does not discriminate reliably between euthyroid and hypothyroid patients and ideally should not be measured in a case of myxoedema.**

D/D of myxoedema :

1. Ageing.
2. Nephrotic syndrome.
3. Angioneurotic oedema.
4. Post menopausal syndrome.

How to treat this case ?

50 ug¹¹ increments at 2-3 weeks interval until an euthyroid state is attained and the dose is then maintained. In majority, usually the maintenance dose is 100-150 ng/day. The drug is given as a

long. Efficacy of replacement therapy is measured by serum TSH level.

If the patient suffers from angina pectoris, start the dose with 25 ug and increase by 25 ug at 4 weeks interval with performing serial ECGs. Increased dose may precipitate anginal pain. During pregnancy, an increase in T₄ dosage about 25-50 ng is often needed.

How to treat myxoedema coma ?

It is a medical emergency and should be treated promptly.

1. L¹⁰ 200 ug IV followed by 20 Mg every 8 hourly, I.V until there is sustained clinical improvement (thyroxine is usually not available for parenteral use; if available, T₄ is given as I.V bolus in a dose of 400-500 ng). If I.V preparation of T is unavailable T may be given 300 ug stat orally and followed by 100-300 ng daily.
2. Hydrocortisone hemisuccinate—100 mg, I.V, 8 hourly.
3. I.V saline infusion—To combat hypotension; monitor cardiac output.

4. **room** **or** hypothermia. Slow swarming of the patient by wrapping with a blanket in a warm

5. High-flow oxygen or assisted ventilation. Correct hypoglycaemia and electrolytes imbalance.
 - treatment of arrhythmias and heart failure accordingly, preferably in an ICU.
7. Antibiotics, as infection is very often the precipitating factor

Recovery within 24 hours is usual. Lifelong T₄ replacement is required after recovery.

Identification points :

eyelids. Tends to have "edematous, swollen" appearance

passionless face with baggy lower

Lh **the 'endocrine system' comprises of 1. Hypothalamus and pituitary 2. Thyroid gland 3 Parathyroid**

Xt "doSL7cX CreaS ^ Adrenal gland's

tissue or mass

Case 70

HYPERTHYROIDISM

Common causes of hyperthyroidism (thyrotoxicosis) :

1. **Graves' disease (diffuse toxic goitre).**
2. **Solitary toxic nodule (adenoma).**

3. Toxic multinodular goitre.
4. Subacute thyroiditis.
5. TSH producing pituitary adenoma.

* The word goitre comes from Latin 'gutter' means throat.

Confirmation of the neck swelling as goitre :

Ask the patient to swallow and note whether the swelling moves upwards on swallowing (because of its attachment to the larynx). Neck swellings which move upwards on swallowing are thyroid, thyroglossal cyst, subhyoid bursitis, pretracheal and prelaryngeal lymph nodes fixed to trachea and larynx respectively. Thyroglossal cyst also moves upwards with the protrusion of tongue.

* Normal adult thyroid gland weighs 20-25 gm.

Examination of the thyroid gland :

- (A) **INSPECTION** : Note the position, diffuse or localised nature of swelling, extent, shape, size, overlying skin (dilated veins, thyroidectomy scar, local signs of inflammation) of the swelling which is present in the neck; as a thyroid swelling moves upwards on swallowing, look for the inferior border as soon as the gland moves up.
- (B) **PALPATION from the behind** :
At first, palpation of the thyroid gland should always be done from the behind on a sitting patient by using both hands (specially, the pulps of the fingers) with slight flexion of the neck (to relax the sternomastoid muscles), and thumbs placed over the nape of the neck. Note the size, shape, consistency, mobility etc.
- (C) **PALPATION from the front** :
 Expose the neck properly and extend the neck slightly. Often each lobe of the thyroid gland is better palpated from the front by **Lahey's method** e.g., for palpation of the right lobe, the gland is pushed to the right from the left side; the right lobe becomes more prominent and now palpated by the left hand. Henceforth, the following points are noted carefully :
 - a) Size - Small or large.
 - b) Shape - Localised or diffuse swelling, nodular or not.
 - c) Surface - Smooth or irregular.
 - d) Consistency - The different consistency are : soft (normal gland), firm (simple goitre), hard (carcinoma of thyroid) or woody feel (Reidel's thyroiditis); a cystic swelling may feel solid and a solid one cystic.
 - e) To get below the swelling - Ask the patient to swallow again (to test the possibility of retrosternal extension).
 - f) Mobility - Fixed or mobile (the gland is moved horizontally and vertically). Mobility is lost in carcinoma of thyroid with infiltration.
 - g) Tenderness - Tender (thyroiditis, bleeding within cyst) or non-tender.
 - h) Relation with the neighbouring structures (pressure effect, fixity etc) ;
 - (i) Trachea - Deviated or not.
 - (ii) Carotid sheath - Carotid pulsation is increased in Graves' disease whereas the pulsation may not be felt in a patient with thyroid carcinoma.
 - (iii) Oesophagus - C/O dysphagia.
 - (iv) Recurrent laryngeal nerve - May produce hoarseness of voice.
 - (v) Sympathetic trunk - Produces Horner's syndrome (pseudoptosis, miosis, anhidrosis of ipsilateral face, and enophthalmos).
 - (vi) Sternomastoid muscle.
 - (vii) Skin and subcutaneous tissue.
 - i) Thrill - Present over the thyroid gland (present in Graves' disease due to increased vascularity).
- (D) **PERCUSSION** : Though not very helpful, percussion of the sternum may be done to detect the presence of retrosternal goitre (read the section on 'Percussion of the sternum or base of the heart'). Changing the percussion note from resonant to dull indicates the possibility of retrosternal goitre.
- (E) **AUSCULTATION** : Search for systolic bruit over the thyroid gland (present in Graves' disease). Ask the patient to hold his breath and then auscultate. Presence of systolic bruit in the thyroid gland may be confused with carotid bruit or venous hum.
- (F) **MEASUREMENT** : Measure the circumference of the neck by a tape at the most prominent part.

* The isthmus which lies in front of 2nd, 3rd and 4th tracheal rings, is better palpated from the front.

**** Thyroid bruit**—more prominent in upper part of neck than lower part (as superior thyroid artery arises from external carotid artery); **Carotid bruit**—obviously more prominent in lower part of neck (over common carotid artery) than upper part; **Venous hum**—disappears on gentle pressure applied over base of the neck.

***** Hyperthyroidism is often associated with goitre (diffuse or nodular).**

******** Cervical lymph nodes should always be palpated to exclude metastasis from thyroid carcinoma.

Pemberton's sign :

Ask the patient to raise both arms above the shoulders until they touch the ears and to keep them in that position for sometime. Facial congestion (plethora), central cyanosis, respiratory distress with stridor, and venous congestion of neck occurs in patient with retrosternal goitre, and is known as Pemberton's sign; it is due to obstruction of great vessels (by pressure effects of retrosternal goitre on arm raising) at thoracic inlet. This test should not be done routinely.

Why do you say that it is Graves' disease ?

As there is presence of exophthalmos and diffuse goitre, it is a patient of Graves' disease.

What is simple diffuse goitre?

It is goitre with no sign of toxicosis, and the causes are iodine deficiency, puberty goitre and use of goitrogens (drugs like amiodarone, lithium, PAS and vegetables like cabbage, cauliflower, turnips).

Features of ophthalmopathy and dermatopathy ;

Graves' disease is characterised by the classical triad of hyperthyroidism, ophthalmopathy and dermatopathy.

- I. Ophthalmopathy — Read the section on 'Exophthalmos'.
- II. Cutaneous manifestations in Graves' disease :
 1. Skin— Warm, hyperhidrotic and moist hand (due to cutaneous vasodilation); palmar erythema, rarely spider naevi, malar flush.
 2. Hair— Friable, fine and silky; alopecia.
 3. Nails— Separation of the distal margin of the nail from its nail bed (Plummer's nail).
 4. Pretibial myxoedema— The affected area (over the shin) is raised, thickened, hyperpigmented and indurated plaque with "peau d'orange" appearance along with growth of coarse hair.
 5. Pigmentation— Hyper- and hypopigmentation (vitiligo) may be seen.
 6. Thyroid acropachy— Clubbing of the fingers with bony changes resembling hypertrophic osteoarthropathy may accompany the dermal changes in distal extremities.

Physical findings in CVS in Graves' disease :

1. Pulse -
 - (i) Tachycardia (persists even when the patient is sleeping, i.e., high sleeping pulse rate).
 - (ii) High volume collapsing pulse which may be of water-hammer in character (due to wide pulse pressure), capillary pulsation,
 - (iii) Atrial fibrillation (irregularly irregular pulse), ectopics.
 - (iv) Carotid dance and suprasternal pulsation (due to hyperkinetic circulatory state).
 - (v) Other points in pulse — Usually within normal limit.
 2. Signs of congestive cardiac failure (high output failure and often refractory) may be present.
 3. Heart -
 - (i) Apex— Hyperdynamic and present in normal position.
 - (ii) S₁ and P₂ may be loud (due to tachycardia).
 - (iii) Murmur— Systolic murmur at the apex (due to increased blood flow).
 - (iv) Means-Lerman scratch — To-and-fro scratchy sound is heard in the pulmonary area in midsystole (possibly due to rubbing of pleura and pericardium as a result of hyperkinetic circulatory state mimicking pericardial rub). It is rarely found.
- * Common symptoms related to CVS are (i) Palpitation with sweating, (ii) Angina pectoris, and (iii) Dyspnoea due to CCF. *Sleeping pulse rate is important to determine the severity of thyrotoxicosis.*

Gastrointestinal manifestations in Graves' disease :

Loss of weight in spite of normal or increased appetite, hyperdefecation as well as diarrhoea and steatorrhoea, vomiting.

Possible causes of increased sleeping pulse rate :

1. Thyrotoxicosis.
2. Rheumatic carditis.
3. Atrial fibrillation.
4. Pulmonary tuberculosis (active).
5. Atropinised patient.
6. Myocarditis due to any cause.
7. Fever due to any cause.
8. Bleeding (internal).

Three cardinal signs of Graves' disease :

1. Tachycardia,
2. Tremor, and
3. Exophthalmos.

N.B. : *Examine these three signs in all patients with hyperthyroidism.*

* Robert J. Graves (1796-1853) was a physician, Meath Hospital, Dublin, Republic of Ireland.

Fades in thyrotoxicosis :

Better known as 'stare'; an anxious startled look due to lid retraction (agitated or terror face). Exophthalmos and other eye changes like Infrequent blinking may be noted. Moist facial skin with a malar flush is sometimes seen and may be associated with a goitre in the neck.

Clinical features regarding involvement of nervous system :**(A) Symptoms-**

1. Insomnia, nervousness, emotional lability or hyperirritability; perspiration.
2. Hyperkinesia.
3. Depression, mania or psychosis.
4. Difficulty in rising from squatting position or climbing upstairs (due to proximal muscle weakness).
5. Stupor or coma (thyroid storm).

(B) Signs-

1. Fine tremor (most prominent feature) - Tested in outstretched hands and in protruded tongue. In severe cases, the whole body may be shaking and trembling.
2. Proximal muscular weakness and wasting - 'Thyroid myopathy'.
3. Myasthenic syndrome and hypokalaemic periodic paralysis may occur.
4. Chorea.
5. Deep reflexes - Exaggerated.
6. Pseudoclonus or ill-sustained clonus.
7. Plantar response - Flexor.

Differential diagnosis of thyrotoxicosis :

(A) Anxiety neurosis is the commonest differential diagnosis (having tremor in both) :

	Anxiety neurosis	Thyrotoxicosis
1. Appetite	1. Diminished	1. Increased (but there is weight loss)
2. Hand	2. Cold and moist	2. Warm and moist
3. Sleeping pulse rate	3. Normal	3. Raised
4. Pulse pressure	4. Normal	4. Increased
5. Goitre	5. Not present	5. Present
6. Eye signs	6. Absent	6. Present

(B) Other conditions where there is rapid weight loss are :

1. Diabetes mellitus.
2. Pulmonary tuberculosis.
3. Malabsorption syndrome.
4. Malignancy.
5. Pheochromocytoma.
6. AIDS.

(C) Chronic alcoholism — Tremor, tachycardia, sweating, emotional lability in alcoholism often resemble thyrotoxicosis.

Examination of *neck' in clinical medicine :

1. Swelling like branchial cyst, sternomastoid tumour, cystic hygroma, carotid body tumour, Ludwig's angina, dermoid cyst etc. are of surgical interest.
2. Skin—Scar, old tracheostomy mark, signs of inflammation, fistula (branchial), sinus, actinomycosis (rare), pityriasis versicolor, Casal's collar, acanthosis nigricans, acrochordons etc.
3. Enlargement of the thyroid gland (page-401).
4. Lymph nodes (page-223).
5. Salivary glands.
6. Neck vessels (page-345).
7. Examination of trachea (page-75); tracheal tug (page-49).
8. Neck rigidity (page-152).
9. Short neck (associated with low hair line) is found in cranio-vertebral anomaly, e.g., platybasia, Klippel-Feil anomaly (page-169).
10. Great auricular nerve—thickened in tuberculoid leprosy (by turning the head to the opposite side, the nerve stands out).
11. Spasmodic torticollis (page-466) or other abnormal movements (e.g., tics).
12. Suprasternal pulsation, carotid dance (page-39 and 41).
13. Webbing of neck in Turner's syndrome.
14. Swelling of neck in conditions like SVC obstruction, subcutaneous emphysema, bull-neck (malignant diphtheria).
15. Deglutition (to confirm a thyroid swelling).
16. Power testing of neck muscles e.g., sternomastoids (page-471); wasting of sternomastoid muscle in myotonic dystrophy.
17. Restriction of neck movement along with pain (flexion, extension, rotation, side-bending) occurs in cervical spondylosis, ankylosing spondylitis, atlanto-axial dislocation.
18. Ciliospinal reflex (page-448).
19. Auscultation—thyroid bruit, carotid bruit, venous hum (page-402); auscultation over trachea.

* First palpate the back and sides of neck, which are better palpated from the front; then the patient is allowed to sit on a stool and the examination (i.e., palpation) is carried out from behind.

Mention the investigations you like to perform In hyperthyroidism :

1. Serum T_3 and T_4 (free and total both) are increased. TSH is very low (< 0.05 μ U/ml or may be undetectable).
2. Radioactive Iodine (I^{131} or I^{132}) uptake (RAIU) is increased.
3. Resin T_3 uptake (RT_3U) is increased.
4. Free thyroxine index (FTI) is increased ($FTI = \text{serum } T_4 \times RT_3U$).
5. T_3 suppression test - After the administration of T_3 , the iodine uptake is not suppressed.
6. Serum TSH level after TRH administration - Blunted response (very helpful test).
7. ECG - Shows sinus tachycardia, ectopics, and / or atrial fibrillation.
8. Thyroid (radionuclide) scan - Done by technetium 99m which shows localised or generalised increased uptake. It is useful in suspected cases of functioning thyroid nodule.
9. Others -
 - (i) Anti-thyroglobulin antibodies—Raised (also elevated in Hashimoto's thyroiditis).
 - (ii) BMR is increased (normal BMR in male is 40 calorie / sq. meter of body surface / hour).
 - (iii) Serum PBI is increased (normal level is 3.6 - 8.5 (ig/dl) — not used now-a-days.
 - (iv) Low serum cholesterol.
 - (v) Serum LATS (long acting thyroid stimulator) is increased—not routinely used.
 - vi) Antibodies against thyroid peroxidase (TPO) enzyme and thyroglobulin are present in most cases of Graves' disease.
 - (vii) Measurement of TSH receptor antibodies (TRAb) to determine the aetiology.
 - (viii) Needle aspiration of thyroid nodule in selected cases.
 - (ix) High urinary as well as serum calcium and phosphate.
 - (x) Raised SGPT (ALT), alkaline phosphatase, bilirubin; glycosuria ('lag storage').

* In 5% patients of thyrotoxicosis (T_3 toxicosis) serum T_3 is much raised but T_4 remains in the upper limit of normal range.

Table 31 : Clinical differentiation between primary (Graves' disease) and secondary thyrotoxicosis

Features	Graves' disease	Toxic nodular goitre
1. Age	1. < 35 years	1. > 35 years.
2. Goitre	2. Smooth, diffuse enlargement	2. Nodular
3. Onset of symptoms	3. With goitre	3. Goitre first, symptoms later
4. Eye signs	4. Present	4. Absent
5. Cardiac symptoms	5. Slight	5. Severe
6. Neuromuscular symptoms	6. Prominent	6. Slight

Outline of management of thyrotoxicosis :

1. **Antithyroid drugs** - Propylthiouracil (100-200 mg, 6-8 hourly), carbimazole, methimazole, iodide or potassium perchlorate is used. While diminishing the toxicity, these drugs increase the size and vascularity of the thyroid gland. Carbimazole is the commonly used drug.
2. **Radioactive Iodine (I^{131})** —Usually 5-10 mCi of I^{131} is given orally; maybe repeated at 12 weeks.

Indications -

- a) All patients > 40 years of age.
- b) Toxicity due to drugs or relapse after surgery.
- c) Thyrotoxic heart disease.

Contraindications -

- a) During pregnancy and lactation (antithyroid drug is the best choice).
- b) In children.

3. Surgery (subtotal or near-total thyroidectomy).

4. General measures -

(i) Good nutritious diet.

(ii) **Propranolol** - Dosage upto 160 mg / day may be given. Propranolol antagonises the sympathomimetic manifestations and thus, it alleviates palpitation, tremor, sweating and lid retraction. Propranolol is contraindicated in CCF, heart block, bronchial asthma, peripheral vascular disease and diabetes.

5. For thyroid eye disease -

(i) Low salt diet and diuretics, with elevation of the head end at night to reduce orbital oedema. Smoking should be stopped.

(ii) Tinted glass with side frames (to protect the eyes from sun, wind and dust) to reduce lacrimation.

(iii) 1% methyl cellulose and hypromellose eye drops for lubrication.

(iv) Prednisolone — 40-80 mg / day with or without cyclosporin in malignant exophthalmos.

(v) External irradiation or lateral tarsorrhaphy to prevent exposure keratitis.

(vi) Orbital decompression or corrective eye muscle surgery, rarely.

6. For dermopathy (pretibial myxoedema) -

(i) Hyaluronidase or triamcinolone (intralesional injection).

(ii) Topical high potency corticosteroids with occlusive dressing.

(iii) Ultra-violet radiation.

(iv) Octreotide is being tried recently.

* Exophthalmos does not necessarily decrease after successful treatment of thyrotoxicosis.

How carbimazole is given ?

The usual dose is 10-20 mg, orally 2-3 times daily till the patient becomes euthyroid. Usually the patient becomes euthyroid in 6-8 weeks and the maintenance dose is then started as 10-15 mg/day for 12-18 months. The maintenance dose is determined by the measurement of T_4 and TSH.

Side effects — Agranulocytosis (patient complains of sore throat), drug rash, drug sensitivity, hepatitis, drug fever, arthralgia etc.

* Leucopenia or agranulocytosis is the most dangerous complication of antithyroid drugs. If the absolute neutrophil count is $<1500 / \text{mm}^3$ of blood, antithyroid medication should be discontinued.

Use of beta-blockers in internal medicine :

Remember the indications from head to foot as,

1. Anxiety states.
2. Migraine (as prophylaxis).
3. Glaucoma (chronic open-angle).
4. Thyrotoxicosis.
5. Prevention of variceal haemorrhage (i.e., portal hypertension).
6. Cardiovascular system— Systemic hypertension, angina pectoris (stable and unstable), myocardial infarction, cardiac arrhythmias (specially tachyarrhythmias), hypertrophic obstructive cardiomyopathy (HOCM), mitral valve prolapse syndrome (MVPS), dissection of aorta, cyanotic spells of Fallot's tetralogy.
7. Essential tremor.
8. Pheochromocytoma.
9. Alcohol withdrawal syndrome.

Thyroid storm or thyrotoxic crisis :

It is an extreme degree of hyperthyroid state precipitated by surgery, infection, severe stress, or develops spontaneously in patients with hyperthyroidism. The patients usually suffer from restlessness, delusions, hyperpyrexia, delirium, tachyarrhythmias, hypotension, vomiting, diarrhoea and often coma, and may succumb from heart failure, arrhythmias and hyperpyrexia..

Identification points (thyrotoxicosis) :

1. Goitre with exophthalmos is the first clue—the patient looks anxious, restless and fidgety.
2. Nov/ examine for tremor and tachycardia.
3. Never forget to examine the jerks.

* **If thyrotoxicosis is given as a long case**, examine the patient in the following way :

- (A) General survey (specially nutrition, facies, neck glands, pulse, BP, skin and appendages).
- (B) Examination of the thyroid gland and eye.
- (C) Systemic examination :
 - a) CVS,
 - b) Nervous system,
 - c) G.I. system,
 - d) Respiratory system, and
 - e) Reticulo-endothelial system.

' ' **Hyperthyroidism** indicates glandular hypertrophy of thyroid; **thyrotoxicosis** is increased circulating free thyroid hormones along with toxic features.

*** Heat intolerance is a common general symptom of hyperthyroidism.

Endocrinal emergencies in clinical practice are thyroid storm, myxoedema coma, pituitary apoplexy, ectopic ACTH syndrome, Nelson's syndrome. Read the section on 'Diabetes mellitus' for diabetic emergencies.

Case 71

EXOPHTHALMOS

What is exophthalmos ?

It is the forward protrusion of the eyeball when a portion of the sclera is visible above and below the cornea. Previously, it was defined as the visibility of the **lower sclera** of more than 2 mm when the patient looks straight forward (few clinicians still believe this definition).

In exophthalmos (unilateral), the globes appear asymmetric.

How the exophthalmos is tested at the bedside ?

1. The patient is asked to look straight forward. The examiner stands **in front of the patient** and the visibility of the upper and lower sclera is noted. **The visibility of the lower sclera**

is more important as in thyrotoxicosis, the presence of lid retraction (contraction of Muller's muscle), may give the false impression of exophthalmos.

2. The examiner stands **behind the patient**, tilts the patient's head backwards and looks vertically down the slanting forehead in the plane of superciliary ridges. Thus, one may be sure whether any part of the eyeball (globe) is visible (proptosis) or not. This is known as **Naffziger's sign**.

* According to Jew clinicians, *exophthalmos* and *proptosis* carry the same message.

** In health, one-third of the cornea is covered by the upper eyelid.

What are the common causes of exophthalmos ?

(I) Unilateral :

1. Early thyrotoxicosis.
2. Retrobulbar tumour.
3. Cavernous sinus thrombosis.
4. Chloroma (AML); malignant and lymphomatous deposits.
5. Orbital 'pseudotumour'.
6. Varicosity of orbital vein, carotico-cavernous fistula, aneurysm of orbital vessels, orbital haemorrhage.
7. Orbital cellulitis.

* So, both eyes are not necessarily affected simultaneously in exophthalmos.

(II) Bilateral :

1. Thyrotoxicosis.
2. Bronchial asthma (long standing).
3. SVC syndrome.
4. Cirrhosis of liver (few cases).
5. Cushing's syndrome.
6. Craniostenosis (e.g., oxycephaly).
7. Cavernous sinus thrombosis (bilateral).
8. Hand-Schuller-Christian disease, nasopharyngeal carcinoma affecting both the orbits.

N.B. : Commonest cause of exophthalmos is thyrotoxicosis whether it is unilateral or bilateral; 2, 4 and 5 of (II) produce apparent exophthalmos due to lid retraction.

* Cavernous sinus thrombosis = chemosis of conjunctiva, exophthalmos, ptosis, ophthalmoplegia, papilloedema and reduced sensation in ophthalmic division of trigeminal nerve.

** Intermittent proptosis : orbital varix, recurrent orbital haemorrhage, periodic orbital oedema.

*** Pseudoproptosis : shallow orbits, pathological myopia and contralateral enophthalmos.

Other eye signs in thyrotoxicosis :

Eye signs in Graves' disease are divided into two parts : 1) Spastic (staring look, lid lag, lid retraction) and 2) Mechanical (proptosis, ophthalmoplegia and congestive oculopathy).

1. Widening of the palpebral fissure.
2. **von Graefe's sign** (lid lag) — When the patient is asked to look downwards suddenly, the upper eyelid lags behind (exposing the sclera between the upper eyelid and cornea).
3. **Joffroy's sign** — Absence of wrinkling on the forehead on looking upwards (head must be fixed).
4. **Moebius's sign** — Lack of convergence of the eyeball (the test for accommodation is done and Moebius's sign is elicited).
5. **Stellwag's sign** (staring look) — Infrequent blinking.
6. **Dalrymple's sign** (lid retraction) — This is equivalent to visibility of the upper sclera when the patient looks forward.
7. **Jendrassik's sign**—Paralysis of extraocular muscles.

* Bilaterally dilated pupil may be found as a result of sympathetic overactivity.

** Swollen extraocular muscles (typical of Graves' disease) and other features of thyroid eye disease are detected by orbital USG or CT scan. The development of unilateral proptosis usually implies a space-occupying lesion and warrants urgent CT scan or MRI of orbit.

Ocular complaints in Graves' disease :

1. Excessive lacrimation.
2. **Grittiness or pain in eyes.**
3. Redness of eye.
4. Photophobia.
5. Diplopia.
6. **Ophthalmoplegia.**
7. Squint.
- g Loss of visual acuity.

Table 32 : Classification of eye changes in Graves' disease

This classification was done by American Thyroid Association. Remember the acronym 'No SPECS'.

Class	Mnemonics	Definition
0	(N)	No signs, no symptoms
1	(O)	Only signs (lid retraction, lid lag), no symptoms
2	(S)	Soft tissue involvement (symptoms and signs)
3	(P)	Proptosis > 22 mm (measured by Hertel exophthalmometer)
4	(E)	Extraocular muscle involvement (external ophthalmoplegia e.g., having diplopia)
5	(C)	Corneal involvement (ulceration)
6	(S)	Sight loss (optic nerve involvement)

Extraocular muscles : earliest affection — inferior rectus, least affection—lateral rectus

Causes of pulsating exophthalmos :

Carotico-cavernous fistula and saccular aneurysm of ophthalmic artery.

Physical examination in exophthalmos (from thyrotoxicosis) ;

1. Warm and moist hand.
2. Pulse rate - Whether tachycardia present or not.
3. Tremor - Ask the patient to stretch out the arms and to spread the fingers. Look for the presence of fine tremor. One may examine the protruding tongue for fine tremor.
4. Goitre present or not (also examine for the presence of systolic thrill and / or bruit present over thyroid gland).
5. CVS - See the section on 'Hyperthyroidism'.
6. Jerks - Exaggerated.

* One should examine these 6 points in all patients with exophthalmos. Features like warm and moist hand, tachycardia, tremor, exaggerated jerks and goitre are evident in Graves' disease.

What is exophthalmic ophthalmoplegia ?

It is the presence of exophthalmos, chemosis of conjunctiva (oedema of conjunctiva and injection of sclera) and weakness of extraocular muscles (sparing pupillary and ciliary muscles) leading to diplopia and squint.

Pathophysiology : Proliferation of fibroblasts within the orbit -> secretion of glycosaminoglycans (hydrophilic) -> increase in interstitial fluid volume -> associated infiltration of chronic inflammatory cells swelling of the extraocular muscles increase in retrobulbar pressure.

What is malignant exophthalmos ?

Progressive bulging of the eyeballs, conjunctival oedema, exposure keratitis with corneal ulceration and visual loss are features of malignant exophthalmos.

What is enophthalmos ?

Ir[^]^h[^]S condition' the eyeball is retracted inwards or recessed within the orbit. It is very rarely seen. The common causes are,

1. Phthisis bulbi (shrinkage of the eyeball from prolonged endophthalmitis) or microphthalmos.
2. Horner's syndrome.
3. Old age (due to atrophy of orbital fat) - physiological; trauma, emaciation.
4. As a consequence of resolved orbital cellulitis.
5. Severe dehydration (facies Hippocraticus).
6. Fracture of the orbital floor.

* Management of ophthalmopathy has been described in the section on 'Hyperthyroidism'.

Case 72

CUSHING'S SYNDROME

What is Cushing's syndrome ?

It is a syndrome with symptom complexes and signs, produced due to prolonged inappropriate elevation of free corticosteroid in the circulation. Commonly it is due to therapeutic administration of steroids or ACTH (iatrogenic). Non-iatrogenic (i.e., Cushing's disease or adrenal adenoma) Cushing's syndrome is rare.

* Harvey Cushing (1869-1939) was Professor of Surgery, Harvard University, Massachusetts, USA.

Diagnosis of the case by inspection :

The disease is four times more common in women than men.

1. Moon face (dusky and plethoric too; excess hair in face).
2. Obesity (truncal obesity with thin limbs).
3. Buffalo hump.
4. Purple striae.
5. Proximal myopathy.
6. Hirsutism in females.

Describe the clinical features of Cushing's syndrome :

- I. Fat deposition -
 - a) Obesity - Weight gain is the commonest symptom and thus, obesity is the most frequent sign. There is centripetal (like a lemon on matchstick) deposition of adipose tissue though generalised obesity is not uncommon. Fat deposition in the mesenteric bed produces 'truncal obesity'. The fat deposition spares the extremities.
 - b) Moon facies - Due to fat deposition in the upper part of the face (supramalar region).
 - c) Buffalo hump - Due to deposition of fat in the interscapular area (upper part).
- II. Skin and hair changes -
 - a) Plethoric appearance - Due to thinning of the skin and rarely due to associated polycythemia.
 - b) *Purple* striae - Commonly seen over abdomen, buttocks, breast, thigh and arms. This is produced due to break-down of collagen fibres in the dermis and thus, the vascularised subcutaneous tissues are exposed. These 'striae distensae' are often painful.
[*White* striae (striae albicantes) are usually observed following pregnancy, after paracentesis abdominis in ascites or when an obese person loses weight rapidly; *pink* striae are common during pregnancy and in simple obesity with rapid increase in weight].
 - c) Easy bruisability and ecchymosis - Due to loss of perivascular supporting tissue (increased capillary fragility).
 - d) Hyperpigmentation - This is due to excess MSH secretion found in bilateral adrenal hyperplasia secondary to pituitary tumour or ectopic ACTH secretion from non-endocrine tumours like bronchogenic carcinoma. This is not a very common feature.
 - e) Thinning of hair.
 - f) Increased incidence of skin infection (e.g., tinea versicolor).
- III. Myopathy - Loss of protein in muscle leads to proximal myopathy (difficulty in combing hairs or getting up from squatting position).
- IV. Bony changes - Osteoporosis is commonly seen in spine, ribs and femurs (due to loss of protein matrix of the bones); patient may present with backache (vertebral compression fracture).
- V. Hypertension - It is due to the increased plasma volume and enhanced vascular response to catecholamines. This appears not to relate to sodium retention as previously believed. There may be ankle oedema or cardiomegaly.
- VI. Diabetes mellitus - Due to antagonistic action of cortisol on insulin.
- VII. Secondary sexual characters -
 - a) Female - Hirsutism, acne, oligomenorrhoea or amenorrhoea, recession of temporal hair, clitoromegaly.
 - b) Male - Loss of libido, impotence may be seen.
- VIII. Psychiatric abnormality -
 - a) Depression.

- b) Emotional instability.
- c) Irritability.
- IX. Growth -
 - a) Growth retardation is a rule in children.
 - b) Diminution of height in adult patients may occur due to collapse of the vertebrae (as a result of severe osteoporosis).
- X. Miscellaneous -
 - a) Cataract.
 - b) Peptic ulcer.
 - c) Poor wound healing with little inflammatory response.
 - d) Mild bilateral exophthalmos.
 - e) Electrolyte disturbance (T Na⁺, 4- K⁺, J, Cl⁻),
 - f) Avascular necrosis of bone.

Causes of moon face :

Read the section on 'Moon face'.

What is hirsutism, virilisation and defeminisation ?

- (A) **Hirsutism** - It is the growth of terminal hair in women in a pattern characteristic of men.
- (B) **Virilisation** (i.e., androgen excess in females)- The characteristic features are :
 - (i) Frontal baldness.
 - (ii) Increase in size of shoulder girdle muscles.
 - (iii) Coarsening of voice.
 - (iv) Acne and seborrhoea.
 - (v) Hirsutism,
 - (vi) Clitoromegaly.
 - (vii) Increased libido.
- (C) **Defeminisation** -
 - (i) Reduction in breast size.
 - (ii) Loss of female body contours.
 - (iii) Amenorrhoea.

Causes of hirsutism :

1. Familial.
2. Idiopathic or simple hirsutism.
3. Cushing's syndrome, virilising ovarian tumour (e.g., arrhenoblastoma).
4. Polycystic ovarian disease (PCOD).
5. Adrenal androgen secreting tumour (adenoma, carcinoma), congenital adrenal hyperplasia.
6. Hyperprolactinaemia.
7. Acromegaly.
8. Drug-induced— Phenytoin, OC pills, androgens, diazoxide, cyclosporin, psoralens, minoxidil.
9. Menopause.

* Hirsutism is divided into two groups (i) with virilisation (severe PCOD, congenital adrenal hyperplasia, ovarian neoplasm), and (ii) without virilisation (other causes).

** Therapy is done by bleaching, waxing, electrolysis, laser, anti-androgens and treatment of cause.

What is Cushing's disease ?

It is the hypercortisolemia producing bilateral adrenal hyperplasia due to excess ACTH secretion from the anterior pituitary gland.

Hormones secreted from adrenal gland :

Adrenal glands comprise of an outer cortex and an inner medulla. The cortex is a part of hypothalamic-pituitary-adrenal axis, which secrete cortisol and adrenal androgen after getting signal from above. The cortex is divided into 3 zones.

- (A) Cortex —
 - ® Zona glomerulosa—mainly secretes aldosterone (mineralocorticoids)
 - ® Zona fasciculata—predominantly secretes glucocorticoids
 - Zona reticularis—mainly secretes sex steroids (androgen, oestrogen and progesterone)
- (B) Medulla —
 - 9 Produces catecholamines (adrenaline, nor-adrenaline and dopamine)

How do you like to investigate your case ?

1. Blood examination - Mild neutrophilic leucocytosis, eosinopenia and lymphocytopenia. Haematocrit may be high.

2. Electrolytes - Hypernatraemia, hypokalaemia, hypochloraemia, metabolic alkalosis.
3. Urine - Glucose may be present in urine and there is elevated urinary 17-hydroxycorticoids.
4. Glucose tolerance test - Impaired glucose tolerance. Diabetes is seen in < 20% patients.
5. X-rays :
 - a) Spine - Osteoporosis in spine produces cod-fish vertebra or fish-mouth like appearance of intervertebral space. Osteoporosis is also seen in pelvis and skull.
 - b) Chest - There may be superior mediastinal widening due to,
 - (i) Bronchogenic carcinoma (produces ectopic ACTH) or bronchial carcinoid.
 - (ii) Fat accumulation, or
 - (iii) Tumour of thymus (may be associated with Cushing's syndrome).
 - c) Skull - Enlargement of the pituitary fossa in Cushing's disease only.
6. CT or MRI scan of the abdomen (adrenal adenoma or carcinoma), brain (pituitary tumour) or chest.
7. Selective adrenal arteriography and venography - Not used now-a-days.
8. 19- [13I] iodocholesterol scanning - Differentiates adrenal tumours from bilateral adrenal hyperplasia.
9. Plasma cortisol level - The normal blood level is 5-25 (ig/dl at 8 A.M. Normally, the cortisol level is lowest at 12 midnight but in Cushing's syndrome the level remains uniformly elevated throughout the 24-hours period (loss of the circadian rhythm).
10. Plasma ACTH level (at 8 A.M.) -
 - (i) Very high level (> 300 ng/1) - Ectopic ACTH secretion due to malignancy (from small-cell carcinoma of the bronchus).
 - (ii) Moderate level (80-300 ng/1) - Ectopic ACTH secretion from bronchial carcinoid (benign tumour) or pituitary dependent disease.
 - (iii) Normal level (10-80 ng/1) or low level - Adrenal disease.
11. 48-hours low dose dexamethasone suppression test - This test is done in two stages. There is failure of complete suppression in Cushing's syndrome. This is a highly sensitive test.
12. Metyrapone test (exaggerated response is seen in Cushing's disease and the test is negative in Cushing's syndrome).
13. Insulin tolerance test — Insulin-induced hypoglycaemia does not stimulate the plasma cortisol level in Cushing's syndrome, which is a feature in normal healthy persons.
14. 24-hours urinary free cortisol excretion > 250 (ig/day is diagnostic (normal value < 90 |ag/day).
15. Demonstration of the adrenal tumour may be done by IVP, aortogram, retroperitoneal air insufflation (not practised now-a-days).

N.B. : i) Adrenal hyperplasia and ectopic ACTH syndrome are ACTH-dependent.

ii) Adrenal adenoma and carcinoma are not ACTH-dependent.

Differentiation of Cushing's syndrome from ectopic ACTH syndrome :

Ectopic ACTH syndrome will have :

- | | |
|--------------------------|---|
| 1. Sudden onset. | 4. Hypertension (more common). |
| 2. Hyperpigmentation. | 5. Hypokalaemic alkalosis (more common). |
| 3. Oedema (more common). | 6. Markedly elevated ACTH level (> 300 ng/1). |

What is 'pseudo-Cushing's syndrome'?

These patients often physically mimic Cushing's syndrome (Cushingoid appearance) and may have high urinary output of 17-hydroxycorticoids. They are : 1. Primary obesity, 2. Chronic alcoholism, and 3. Severe depression.

Management of your case :

I. Adrenal adenoma :

Surgically removed via a loin approach and suboptimal replacement therapy with 0.5 mg of dexamethasone is started for a variable length of time.

II. Adrenal carcinoma :

Resected as far as possible and the tumour bed is irradiated. Treatment with O, P'-DDD (2-3 g/day) is started immediately.

III. Cushing's disease :

- (i) Trans-sphenoidal removal of the adenoma (treatment of choice).
- (ii) Radical hypophysectomy - If no tumour is seen.

(Iii) Bilateral adrenalectomy with pituitary irradiation with yttrium-90 implantation to prevent the development of Nelson's syndrome.

IV. Ectopic ACTH secretion :

- (i) Surgical removal or therapy of primary tumour (small-cell carcinoma of lung; tumour of the pancreas, thymus, ovary; bronchial carcinoid etc.)
- (ii) 'MEDICAL OR CHEMICAL ADRENALECTOMY' is done by—
 - a) Metyrapone (2-3 g/day).
 - b) O, P'-DDD or mitotane (2-3 g/day).
 - c) Aminoglutethimide (1 g/day).
 - d) Ketoconazole (600-1200 mg/day).

* Mifepristone is a new treatment option for medical adrenalectomy.

V. Supportive therapy is done by :

- (i) High protein intake.
- (ii) Treatment of hypertension, diabetes mellitus, electrolyte imbalance etc.
- (iii) Psychiatric treatment.

* Causes of death are : hypertension, myocardial infarction, heart failure and secondary infections.

What is meant by primary adrenocortical deficiency ?

It is known as Addison's disease (opposite to Cushing's syndrome) and is characterised by,

- (i) Asthenia, weakness and weight loss (glucocorticoid deficiency),
- (ii) Hyperpigmentation of buccal mucous membrane, face, new scars, palmar creases, pressure points (ACTH excess)—dull, slate-coloured, and
- (iii) Hypotension (mineralocorticoid deficiency).

* **i** Cortisol → T ACTH → T melanin → pigmentation; vitiligo (10-20%) may occur due to autoimmunity.

What is 'adrenal crisis' or acute hypoadrenalism ?

(A) It is a symptom complex of anorexia, nausea, vomiting, abdominal pain, fever, somnolence, hypovolaemic shock with low pulse volume and low BP, confusion or coma. It is commonly precipitated by,

- (i) Infection or septicaemia.
- (ii) Surgical stress.
- (iii) Fluid loss (diarrhoea).
- (iv) Sudden withdrawal of long-term corticosteroid therapy.

(B) It may be due to affection of the adrenal gland by,

- (i) Tuberculosis, (iii) Autoimmune destruction, or
- (ii) Metastasis, (iv) Surgical destruction.

(C) Treatment is done by.

- (i) Hydrocortisone hemisuccinate 100 mg, I.V, 6 hourly for a few days.
- (ii) Normal saline and 5% glucose drip — Several litres are required within the first few hours.
- (iii) Dopamine as a vasoconstrictive agent.
- (iv) Treatment of the precipitating cause e.g., antibiotics for infection.
- (v) Mineralocorticoid replacement by fludrocortisone (0.1 mg, orally) after stoppage of saline.

* Bilateral adrenal haemorrhage from meningococcaemia (Waterhouse-Friderichsen syndrome) is a known aetiology.

What is Nelson's syndrome ?

10-20% patients of Cushing's syndrome undergoing bilateral adrenalectomy develop chromophobe adenomas with hyperpigmentation and erosion of the sella turcica in the subsequent years. The plasma ACTH level in these patients are extremely high. This syndrome does not occur if these patients are immediately treated with pituitary irradiation in addition to bilateral adrenalectomy. This condition is treated by surgery, radiotherapy and drugs (sodium valproate).

Classification of glucocorticoid preparations :

- (A) Short-acting (1/2 life is < 12 hours) :
 - (i) Cortisol (hydrocortisone) (ii) Cortisone.
- (B) Intermediate-acting (1/2 life is 12-36 hours) :
 - (i) Prednisone (ii) Prednisolone (iii) Methylprednisolone (iv) Triamcinolone.
- (C) Long-acting (1/2 life is > 48 hours) :
 - (i) Paramethasone (ii) Betamethasone (iii) Dexamethasone.

N.B. : If the potency of cortisol is taken as 1, the potency of prednisolone will be 4 and that of dexamethasone will be 30-40; so dexamethasone is 8-10 times more potent than prednisolone.

Identification points :

1. Obesity.
2. Face :
 - (i) Moon face.
 - (ii) Dusky and plethoric.
 - (iii) Hirsutism.

Case 73

ACROMEGALY

How to diagnose acromegaly by inspection ?

1. Prognathism (lantern jaw or bull-dog jaw) — Lower incisors (mandible) protrude in front of the upper incisors (maxilla), and widened spaces between upper and lower teeth; heavy chin.
2. Enlargement of the face as a whole with prominent supraorbital ridges as a result of enlargement of maxillary, frontal and ethmoid sinuses; frontal bossing.
3. Macroglossia with thick lips, large ears and fleshy nose (tongue may show indentations from the teeth); wide-spacing of teeth.
4. Thick skin with hypertrichosis (e.g., bushy eyebrows), hyperhidrosis and increased sebum production (greasy skin); exaggerated nasolabial folds.
5. 'Spade-shaped' hand (broad hand with thick, large, square-tip fingers) with warm, sweaty and doughy feeling; broad toes.
6. Deep, husky and resonant voice due to increased thickness of vocal cord.

* It is a disease of middle age. Common complaints are headache, changes in appearance, visual-field defects, soft tissue swelling and increased sweating.

** Larger ears and nose are due to cartilaginous overgrowth.

*** Females may have a masculinized appearance with mild hirsutism.

**** Other causes of **prognathism** are ; Fragile-X syndrome, osteopetrosis (certain variety) and nemaline myopathy. In prognathism, lower teeth overbites the upper teeth.

Cranial nerve involvement in acromegaly :

1. Bitemporal hemianopia (pressure on optic chiasma)— Commonest.
2. Blindness due to optic atrophy (affection of optic nerve).
3. External ophthalmoplegia (paralysis of III, IV and VIth cranial nerves).
4. Deafness (VIIIth nerve palsy).

* So, visual field testing should be done in all cases of acromegaly.

** Other 'pressure effects' by the enlarged pituitary gland may be upward pressure to hypothalamus (polyphagia, polydipsia, thermal dysregulation), on temporal lobe (complex partial seizure), and features of raised intracranial tension.

Metabolic and circulatory changes in acromegaly :

1. Glucose intolerance (25%).
2. Diabetes mellitus (10%), and
3. Hypertension (25%).

Investigations you like to perform :

(A) Hormone assay :

1. Radioimmunoassay shows increased basal level of GH (normal level upto 1 mU/L).
2. GH level fails to be suppressed < 1 mU/L during an oral glucose tolerance test (GTT).
3. Estimation of plasma somatomedin C level (i.e., IGF-1) by radioimmunoassay (almost always raised in acromegaly).

* Due to pulsatile nature of GH secretion, a single random GH level is not useful for diagnosis. In 30% patients, the serum prolactin level is elevated due to co-secretion by the tumour. In acromegaly, insulin-like growth factor 1 (IGF-1) levels are increased and the major biochemical effects of growth

hormone (GH) is mediated by IGF-1. Thus, atheromatous disease, colon polyps and carcinoma of colon may complicate acromegaly in the long run.

(B) Visual field examination—a common defect is bitemporal hemianopia.

(C) Radiological changes :

1. Skull (lateral view) -
 - a) Enlargement of the pituitary fossa.
 - b) Sometimes, there is double floor of pituitary fossa.
 - c) Erosion of clinoid process.
 - d) Prominent supraorbital ridges and jaw.
 - e) Enlargement of the frontal and maxillary sinuses.
2. Extremities -
 - a) Tufting of the terminal phalanges, i.e., 'arrow-head appearance'.
 - b) 'Heel-pad thickness' is increased i.e., > 18 mm in women and >21 mm in men.
3. Spine - There is evidences of kyphoscoliosis and osteoporosis.
4. Chest - Cardiomegaly.
5. Ultrasonography of abdomen—Organomegaly (specially hepatosplenomegaly).
6. CT or MRI scan of brain to visualise pituitary gland—Reveals pituitary (macro-) adenoma.

(D) Biochemical :

1. Oral glucose tolerance test - Impaired.
2. Raised serum phosphorus and alkaline phosphatase level.
3. Elevated urinary calcium and hydroxyproline.

* **Heel pad, thickness'** is increased in obesity and oedema-producing states.

Assessment of 'activity' in acromegaly :

1. Increasing size of gloves, rings, hats or shoes.
2. Ill-fitting dentures.
3. Excessive sebum secretion.
4. Excessive sweating.
5. Increasing headache.
6. Increasing visual field defect.
7. Serial photography (gradually progressive macrosomia).
8. Rise in the plasma GH and somatomedin C levels.

What is gigantism ?

Hypersecretion of GH before the fusion of epiphysis is **gigantism**. **Acromegaly** is hypersecretion of GH after the fusion of epiphysis and usually results from a pituitary macroadenoma.

Why the nomenclature 'acromegaly' ?

'Acral' means peripheral (hands and feet), and 'megaly' means enlargement.

What is the opposite terminology of 'prognathism' ?

Micrognathia (receding chin), and is found in Pierre-Robin syndrome and Treacher-Collins syndrome.

Commonest pituitary 'hormone producing' tumour :

Prolactinoma.

Features in respiratory and cardiovascular system in acromegaly :

- Respiratory system—deep and husky voice, increased size of sinuses, obstructive sleep apnoea syndrome.
- CVS—systemic hypertension, cardiomegaly, cardiomyopathy, CCF.

Other features of acromegaly :

- | | |
|---------------------------|--|
| 1. Arthropathy. | 6. Carpal tunnel syndrome. |
| 2. Osteoarthritis. | 7. Visceromegaly (liver, spleen, heart, kidney, tongue). |
| 3. Pseudogout. | 8. Increased skin pigmentation; fibromata mollusca. |
| 4. Myopathy (proximal). | 9. Acanthosis nigricans. |
| 5. Peripheral neuropathy. | 10. Galactorrhoea (in females). |

Causes of carpal tunnel syndrome (CTS) :

1. Rheumatoid arthritis.

2. Myxoedema.
3. Acromegaly.
4. Compression of median nerve by oedema, tenosynovitis, fracture, fibrosis or fascitis.
5. Primary amyloidosis.
6. Pregnancy in the third trimester (due to oedema) or premenstrual oedema.
7. Osteoarthritis, rarely.
8. Exposure to vibration (drivers of tractor, mobile crane; workers involved in chipping, grinding or using drills), eosinophilic fascitis, systemic sclerosis.
9. Diabetes mellitus.
10. Obesity.
11. Patients on haemodialysis (due to deposition of P₂-microglobulin, an amyloid).
12. Idiopathic (probably the commonest cause)—common in middle aged, obese female.

CTS is due to entrapment of median nerve while it passes through the carpal tunnel at wrist. Nocturnal pain, paraesthesia and numbness of radial three-and-a-half fingers are complained, commonly by a female patient. There may be weakness of the abductor pollicis brevis muscle (at thenar eminence) with or without wasting. Tapping over the median nerve at carpal tunnel produces paraesthesia along the cutaneous distribution of the nerve (Tinel's sign), and the symptoms may be reproduced on passive maximal wrist flexion (Phalen's test); raising the BP above systolic pressure for 2 minutes produces paraesthesia (tourniquet test). Nerve conduction study confirms the diagnosis.

A wrist splint at night, a local corticosteroid injection or surgical decompression of the carpal tunnel are different modes of treatment in CTS.

Different 'entrapment neuropathy' in clinical medicine :

- Median nerve (at wrist)—carpal tunnel syndrome.
- Ulnar nerve (at wrist in Guyton's canal).
- Radial nerve (at spiral groove of humerus after a fracture).
- Lateral femoral cutaneous nerve (at lateral thigh)—meralgia paraesthetica.
- Common peroneal nerve (at the neck of fibula).
- Posterior tibial nerve (at the flexor retinaculum at ankle joint)—tarsal tunnel syndrome.

Outline of management of acromegaly :

1. Medical -

Bromocriptine (long-acting dopamine agonist) — Usual dose is 10-60 mg daily in divided doses. It is started in a low dose of 1.25-2.5 mg/day and the dose is gradually increased. Side effects are nausea, vomiting, dyskinesia, postural hypotension, constipation etc. Bromocriptine is useful along with other modalities of therapy. Another dopamine agonist used is cabergoline (0.5 mg daily) which also shrinks tumours. Now-a-days, somatostatin analogues (octreotide or lanreotide) are treatment of choice in resistant cases. The dose of octreotide is 50 ng tds to 1500 ng/day by SC injections, and that of lanreotide is 30 mg I.M depot preparation given daily. Growth hormone receptor antagonists (e.g., pegvisomant; 10-20 mg, SC, daily) are promising drugs.

2. Surgical -

1. Trans-sphenoidal (for small tumours) and trans-frontal surgery (for large tumours).
2. Stereotaxic placement of probes (radio-surgery).
3. Cryosurgery.

3. Radiotherapy -

- a) External gamma irradiation (total 4500 rads).
- b) Accelerated proton beam (total doses upto 10000 rads may be given).
- c) Local implantation of radioactive isotopes of yttrium or gold into the pituitary fossa via a transnasal stereotaxic operation.

* Surgery is usually required in visual field defect. Trans-sphenoidal approach is regarded as first line of treatment; external radiotherapy (gamma knife) is employed as second line of treatment if symptoms persist after surgery. Control of hypertension and diabetes is done accordingly.

** The target of treatment is to achieve a mean GH level < 5 mU/L.

Identification points :

1. Prognathism.
2. Enlargement of the face, and
3. 'Spade-shaped' hand.

Case 74

PALPATION OF BASE OF THE HEART

Method of palpation of base of the heart :

The **patient will sit and lean forward**. Standing on the right side of the patient, put your right palm over the sternum transversely in such a way that your fingers lie over the *pulmonary area*, centre of the palm rests over the sternum, and thenar-hypothenar eminences lie over the aortic area.

* 'Base of the heart' includes aortic and pulmonary area.

What to palpate ?

1. Pulsation.
2. Palpable heart sounds, and
3. Thrill.

Palpate aortic, pulmonary, mitral and tricuspid areas :

- (A) Aortic area—patient will sit and lean forward; put your right palm over right 2nd ICS.
- (B) Pulmonary area—patient will sit and lean forward; put your right palm over left 2nd ICS.
- (C) Mitral area—read the section on 'Mitral incompetence' (i.e., palpate the apical area).
- (D) Tricuspid area—patient will be lying in supine position. Put your right palm over left 4th ICS (close to sternum).

Causes of pulsation over the pulmonary area :

- | | |
|-----------------------------------|---|
| 1. Children (physiological), | 4. PDA, |
| 2. Severe pulmonary hypertension, | 5. VSD, |
| 3. ASD, | 6. Idiopathic dilatation of pulmonary artery. |

Causes of palpable heart sounds in precordium :

For palpation of heart sounds, use whole flat of the right palm.

- (A) Palpable S_1 (mitral area)—produces 'tapping' apex beat in MS.
- (B) Palpable S_2 (P_2)—Best palpable in pulmonary area.

It is known as diastolic shock or 'diastolic knock' and is found in pulmonary hypertension of any aetiology. Read the section on 'Mitral incompetence' for the causes of pulmonary hypertension.

- (C) Palpable S_3 —In LVF (mitral area) or RVF (lower left parasternum).
- (D) Palpable S_4 (mitral area)—responsible for 'double' kicking apex beat in IHSS.

For demonstration of any event (palpation, percussion or auscultation) in aortic or pulmonary area, always ask the patient to sit and lean forward.

Pulsation over the aortic area :

Aneurysm of the arch of aorta.

Causes of thrill in the base :

Read the section on 'Aortic stenosis'.

Palpation for parasternal (left) heave :

Patient lies supine. Place your thenar and hypothenar eminences (heel of the palm), or the ulnar border of the right hand over the lower left parasternum (i.e., 3rd, 4th and 5th ICS). Standing on the right side of the patient, look for any lifting of the hand. The causes of heave are :

- | | |
|---|-------------------------------------|
| a) RVH due to any cause, | c) ASD (left parasternal lift), and |
| b) Left atrial hypertrophy (rare; e.g., in MI), | d) Aortic aneurysm. |

* Heave means the impulse is forceful and it is well sustained; 'lift' means the impulse is forceful but it is ill sustained.

Case 75

PERCUSSION OF THE STERNUM OR BASE OF THE HEART

Three cardinal rules of percussion :

See page 82.

Why percussion is not done routinely in CVS ?

1. Not much informative (has been replaced by chest X-ray and echocardiography).
2. Dislodgement of vegetations may occur in a case of MS, acute myocardial infarction (AMI) or atrial fibrillation. Thus, it may precipitate systemic embolism.
3. It is a cumbersome method in female patients.

Situations where cardiac percussion is still helpful :

1. Pericardial effusion (cardiac dullness is increased),
2. Emphysema (cardiac dullness is lost),
3. Aneurysm of the arch of aorta (left 2nd ICS will be dull on percussion).
4. Dextrocardia.

* There is loss of cardiac dullness in left-sided pneumothorax too.

Percussion note over sternum :

It is resonant because aerated lungs are present behind the sternum.

How the sternum is percussed ?

Sternum may be directly percussed by percussing finger (like the percussion of clavicle) Here, the sternum itself acts as pleximeter finger. Actually in clinical practice, the method described below is followed for percussion of the sternum.

How to percuss the base of the heart ?

1. Ask the patient to sit. Percussion is usually done in the 2nd ICS.
2. First place the pleximeter finger in the aortic area parallel to right sternal border.
3. The line of percussion in the *aortic area* will be perpendicular to right sternal border and go on percussing upto the middle of the sternum (i.e., go from right to left).
4. Now place the pleximeter finger in the *pulmonary area* parallel to left sternal border^ The line of percussion in the pulmonary area will be perpendicular to left sternal border and percuss upto the middle of the sternum where you left (i.e., now go from left to right).
5. One may percuss the aortic and pulmonary areas by the above method and may stop the percussion after reaching right and left borders of the sternum respectively. Then percussion of the sternum may be done directly (alternative method).
6. Listen the percussion note carefully. Thereafter, percussion may be done in the 3rd ICS.

* This total method may be used for percussion of base of the heart or percussion of the sternum or 'mediastinal percussion'.

Dullness over pulmonary area :

1. Aneurysm of the aorta,
2. Massive pericardial effusion,
3. Any mediastinal mass.

Dullness over sternum :

Sternum is dull on percussion due to superior and anterior mediastinal mass like,

- | | |
|----------------------------|------------------------------|
| 1. Lymphoma, | 5- Retrosternal goitre, |
| 2. Bronchogenic carcinoma, | 6. Teratodermoid, |
| 3. Thymoma, | 7- Metastatic carcinoma, and |
| 4. Aortic aneurysm, | 8. Oesophageal cysts. |

N B ■ If a dull note is obtained over the sternum, immediately look for any venous prominence in the neck or swelling of the face and arm, i.e., for the signs of 'SVC syndrome'. SVC syndrome is an indirect evidence of the presence of mediastinal mass. Never forget to percuss the sternum in a patient with generalised lymphadenopathy (e.g., mediastinal adenopathy in lymphoma).

How will you percuss the heart ?

Cardiac percussion is not routinely practised now-a-days for reasons mentioned earlier.

1. At first find out the upper border of liver dullness along the right MCL.
2. Now to choose your space of percussion for the delineation of right border of heart, select 'one space higher' the upper border of liver dullness. Keeping the pleximeter finger parallel to the arbitrary right border of heart, lightly percuss from right to left. Actually percussion is done in the 3rd and 4th ICS. As soon as the dull note is obtained (due to heart), mark it and then join the points. This is the **right border of heart**.

3. Now, localise the apex.
4. For the **left border of heart**, percuss along (or parallel to) the 'left acromio-xiphoid line' (an imaginary line from left-sided tip of the acromion process to the xiphisternum) in the 2nd 3rd and 4th ICS. Join the points of dullness with the apex.
5. Lastly, percuss the base of the heart to delineate the **upper border** (left 2nd and 3rd ICS). Percussion is done parallel to parasternal line (line in between MCL and lateral sternal line)

Normally right border remains retrosternal; if the dullness is parasternal, think of pericardial effusion, aneurysm of the ascending aorta, dextrocardia, mediastinal mass etc. Left border usually remains along the apex beat; if it is outside the apex beat, think of pericardial effusion. *Remember normally the left 2nd ICS is resonant and the left 3rd ICS is dull on percussion-*, if the 2nd ICS gives dull note, think of pericardial effusion, mediastinal mass, aortic aneurysm, pulmonary hypertension.

Cardiac dullness is also increased in cardiomegaly due to any cause (left border will be corresponding to apex). In pericardial effusion, left border will be outside the apex.

N.B. ; In case of loss of either of liver or cardiac dullness, exclude the presence of right- or left-sided pneumothorax respectively; if both the dullness are lost, think of presence of emphysema.

Case 76

AUSCULTATION OF DIFFERENT AREAS OF HEART

Auscultation of the mitral area :

1. First you have to **localise the apex beat** (i.e., the mitral area) by proper method of palpation (read the section on 'Mitral incompetence'). Mitral area corresponds to cardiac apex.
2. Now auscultate the mitral area in dorsal decubitus position and then in **left lateral position** and at the height of **expiration**. Conventionally, **first use the diaphragm** of stethoscope and **then the bell**.
3. In case of a faint diastolic murmur, you may allow the patient to do mild exercise (if the present condition of the patient permits to do so, i.e., if there is no dyspnoea, CCF arrhythmia, cyanosis etc). Allow the patient to sit and touch both feet with both of his hands and lie down consecutively for five to six times. The murmur of MS becomes louder and prominent after mild exercise (before doing the exercise, you may take permission of the examiner).
4. In the presence of mitral systolic (pansystolic) murmur, you may auscult the left axilla and interior angle of left scapula for radiation of MI murmur (always take the permission of the examiner because you are shifting your stethoscope from the mitral area).
5. Comment on murmur, heart sounds (specially, on S₁), any split, click, opening snap or pericardial rub.

If the apex beat could not be localised properly, auscultate the area 'below the left nipple'.

Auscultation of the aortic area :

1. The right 2nd ICS (close to the sternum) is the aortic area.
2. Allow the patient to **sit and lean forward**. Now auscultate the aortic area at the height of **expiration** with the **diaphragm** of stethoscope.
3. If you have to confirm the radiation of murmur to carotids (AS) or towards the neo-aortic area (AI), you have to take the permission of the examiner.
4. Comment on murmur, heart sounds (specially, on A₂) or any other sounds like split, click etc.

Auscultation of the pulmonary area :

1. The left 2nd ICS (close to the sternum) is the pulmonary area.
2. Patient will **sit and lean forward** and auscultation is done with the **diaphragm** of stethoscope, at the height of **inspiration** (as right-sided events are pronounced in inspiration).

Auscultation of the tricuspid area :

1. The left 4th ICS (close to the sternum) is the tricuspid area.
2. Auscultation is done with the **diaphragm** of stethoscope, at the height of **inspiration** with the patient **lying** in supine position.

Importance of the neo-aortic area :

The left 3rd ICS (close to the sternum) is known as neo-aortic area or **Erb's point**. Often the early

diastolic murmur of AI is best audible in the neo-aortic area. The area should be auscultated in **sitting and leaning forward position** with the diaphragm of stethoscope, and at the height of **expiration**.

Points to note during auscultation of CVS :

1. Heart sounds,
2. Murmur,
3. Splitting of heart sounds, ejection click, opening snap, and
4. Others : Pericardial rub, pericardial knock, prosthetic valve sounds, tumour plop.

Sequence of auscultation of heart :

Apex → proceed along the left border of sternum, i.e., tricuspid area → neo-aortic area → pulmonary area → then auscultate aortic area auscultate different areas, with auscultatory manoeuvres, if required.

But majority of clinicians prefer to examine or write in the following way : apex → pulmonary area aortic area → tricuspid area → neo-aortic area.

Points to note in a murmur :

Murmurs are abnormal cardiac sounds produced by turbulent blood flow (laminar flow is distorted to turbulent flow) through the abnormal valves (MS, MI etc), normal valves (pregnancy, severe anaemia), great vessels (any arterial stenosis will produce bruit) or blood passing through an abnormal channel (VSD, ASD etc). The points noted in a murmur are ;

1. Where best heard or point of maximum intensity (enumerate the area).
2. Timing (systole, diastole or continuous).
3. Character (e.g., blowing, harsh, musical, rumbling) and pitch.
4. Radiation or direction of selective propagation (radiation indicates the direction of blood flow).
5. Intensity (grading; if thrill is present, it is grade 4 or 4+).
6. Change with respiration.
7. Change with posture.
8. Change after exercise.
9. Whether best audible with the diaphragm or the bell of stethoscope.

* Always try to describe a murmur with these NINE POINTS.

** 'Pitch' of a murmur is determined by the flow of blood, i.e., low-pitch indicates low-velocity flow and high-pitch indicates high-velocity flow.

*** Systolic murmurs coincide with carotid upstroke and diastolic murmurs precede the carotid upstroke.

Explanation of effects of respiration or posture on murmur :

During inspiration the intrapleural pressure becomes more negative and thus, more blood comes in the right atrium. So, during inspiration there is increase in the stroke output of the right heart and reduction in the stroke output of the left heart. Similarly stroke output of the left heart increases on expiration. So the murmurs originating in the right side of the heart become louder and prominent on inspiration, and that of the left side of the heart become more intense on expiration.

In the left lateral position, the apical area (mitral area) comes closer to the chest wall, so does the base of the heart in sitting and leaning forward position. It is why the murmurs in those areas become more intense on adopting the stated postures.

* Low-pitched sound e.g., murmur of MS and TS are best auscultated by the bell of stethoscope. All other murmurs are best heard with the diaphragm of stethoscope.

Case 77

Pericardial RUB

Characteristics :

It is a leathery sound produced by the rubbing of inflamed visceral and parietal pericardium :

1. Superficial, scratchy or leathery, inconstant or evanescent (varies from hour to hour), high-pitched, to-and-fro sound (mimics the sound of husking machine); 'close to the ear' quality.

2. Usually best heard on the left side of lower sternum (may be heard over the entire precordium)—here lies the bare area of heart i.e., part of heart not covered by the lung.
3. Intensity of the sound increases when patient sits and leans forward (thus, always auscultate in sitting position of the patient).
4. Intensifies by pressing the chest piece (diaphragm) of stethoscope.
5. It may have presystolic, systolic and early diastolic components.
6. **Sound continues even after holding the breath.**
7. May be associated with chest pain.
8. Usually there is no transmission (i.e., localised).
9. At times it is palpable—The 'tactile fremitus' (palpable pericardial rub).

What is your case ?

This patient is suffering from acute dry pericarditis and there is presence of pericardial rub (describe the characteristics). *The hallmark of diagnosis of pericarditis is pericardial rub.*

Common causes of acute pericarditis :

1. Idiopathic (without any obvious cause),
2. Viral (H/O fever, cough etc—commonly Coxsackie B virus infection),
3. Tuberculous (H/O haemoptysis, anorexia, night sweats etc),
4. Pyogenic (high rise of temperature with toxic features),
5. Rheumatic fever (arthritis, pancarditis, subcutaneous nodules),
6. Uraemia (oedema, anaemia, oliguria and usually painless pericarditis),
7. Myxoedema (delayed ankle jerk, hoarseness of voice),
8. Acute myocardial infarction (acute chest pain with collapse) during acute attack, or Dressler's syndrome (after few days to weeks, following an attack of myocardial infarction),
9. Trauma (H/O trauma),
10. Collagen vascular diseases like SLE, systemic sclerosis (skin rash, arthralgia),
11. Neoplastic or post-irradiation (H/O radiation, lymphadenopathy) etc.

* 1,2 and 3 are most common causes, specially in India.

Describe the chest pain in pericarditis :

Pain is an important but not invariable symptom in pericarditis. *Pain is absent* in slowly developing tuberculous, hypothyroid, neoplastic, post-irradiation or uraemic pericarditis.

There are three types of pain :

1. Pleuritic (steady and constrictive), i.e., due to associated pleurisy and caused by direct extension of infection and thus, the pain is aggravated by inspiration, coughing, sneezing, jolting, change of posture etc. The rub is known as 'pleuro-pericardial rub'.
2. Steady constricting retrosternal pain which radiates in either of both arms, shoulders and neck like acute myocardial infarction.
3. Often the pain is present characteristically in the centre of the chest or left precordial, referred to the back, arms and shoulder.

* The pain of pericarditis is due to associated pleurisy or inflammation of the lower 1/3rd of the parietal pericardium (visceral pericardium is pain-insensitive) supplied by phrenic nerve. The pain varies with breathing and posture, and is often relieved by sitting up and leaning forward but increased by lying supine.

Investigations you like to perform :

- (A) Blood—TC, DC, ESR, cholesterol, urea, creatinine, ASO titre, T₄ and TSH, SCOT (AST).
- (B) Mantoux test— For tuberculous aetiology.
- (C) ECG—**ST-segment elevation with concavity upwards**, and flattening or inversion of T-waves.
- (D) Chest X-ray (in upright posture)-
 1. To exclude tuberculosis in the lungs.
 2. To exclude bronchogenic carcinoma or lymphoma (mediastinal widening).
 3. Enlarged cardiac shadow may indicate very early effusion.
- (E) Echocardiogram— May be helpful in detecting thickening of pericardium and early effusion.

* ECG changes of acute myocardial infarction is ST-segment elevation with *convexity* upwards.

D/D of pericardial rub :

- | | | |
|----|---|---|
| 1 | Systolic murmur | 1 consistent in character, audible in all |
| 2 | Early diastolic murmur of aortic incompetence | ^positions, soft/musical/machinery |
| 3. | Continuous murmur | J sound, pain is not associated with |
| 4. | Artefact. | |
| 5. | Hamman's sign. | |

What is Hamman's sign ?

It is a crunching, rhythmical sound heard over the precordium and is synchronous with the heart beat. This sign is found in pneumomediastinum (or mediastinal emphysema). The sound is best audible in left lateral position. Always search for subcutaneous emphysema meticulously.

Changes appearing if pericardial effusion develops :

(A) Symptoms—Patient will C/O heaviness (retrosternal oppression) in the precordium. dyspnoea, palpitation, swelling of feet or whole body, dry cough etc.

(B) Signs—

1. Orthopnoea (sometimes, patient bends forward in sitting position mimicking the position adopted by Mohammedans during prayer).
2. Face—Puffy.
3. Pulse—Tachycardia, low volume, pulsus paradoxus (specially in acutely developing pericardial effusion), other points in pulse are within normal limit.
4. Cyanosis—If present, mainly of **peripheral** in type.
5. Neck veins—Engorged and may be pulsatile; Kussmaul's sign may be present (rare).
6. BP—Systolic BP is low.
7. Oedema—Pitting in nature.
8. Heart—
 - a) Bulging of precordium with intercostal fullness.
 - b) Apical impulse—Neither visible nor palpable.
 - c) Cardiac dullness is increased on percussion. If apex beat is palpable, it will be feeble and will remain within the outer border of cardiac dullness. Percussion note over the sternum and left 2nd ICS is dull. Tympanicity on Traube's space percussion is retained.
 - d) Heart sounds become faint or muffled. Pericardial friction rub is not always abolished.
9. Lungs—No adventitious sound is heard. Presence of Ewart's sign which is,
 - a) Dullness on percussion,
 - b) Increased vocal resonance, and
 - c) Bronchial breath sound (tubular) — All the features are due to compression collapse of the left lung by massive pericardial effusion and is found at the inferior angle of left scapula (i.e., base of left lung).
10. Alimentary system—
 - a) Liver—Enlarged, soft and tender (congestive hepatomegaly).
 - b) Ascites—May be present.
11. ECG—low voltage QRS complexes, electrical alternans (rare).
12. Chest X-ray (PA view)—enlarged cardiac silhouette, globular or 'pear-shaped heart, oligoemic lung fields and positive Rotch's sign. Echocardiography confirms the diagnosis.

* Pulsus paradoxus and Kussmaul's sign are hallmarks of cardiac tamponade. In cardiac tamponade, there is collection of fluid in the pericardial sac in an amount sufficient to produce serious obstruction to inflow of blood to the ventricles and thus, results in acute heart failure (there is decrease in stroke volume due to impaired cardiac contraction). The amount of fluid may be as small as 250 ml when the fluid collects rapidly or it may be more than 1000 ml in slowly developing effusions. It is a medical emergency and pericardiocentesis is life saving.

Uraemic pericarditis : what to do ?

If pericardial rub is heard in a patient of uraemia, immediately send the patient for dialysis.

Treatment of acute pericarditis :

1. Rest in bed.
2. NSAID—indomethacin (25 mg, thrice daily, orally after meal) may be used.

3. Glucocorticoid—Prednisolone 20-80 mg daily in selected cases; does not accelerate cure; only gives symptomatic relief. If there is no response to corticosteroid, azathioprine or colchicine may be tried.
 4. Treatment of the underlying cause (e.g., antibiotics, antituberculosis drugs).
- * The patient is observed frequently for the development of pericardial effusion.

Conclusion :

— ^ In a suspected case with pericardial rub, ask the patient to hold the respiration. One should do this to differentiate pericardial rub from left-sided pleural rub. **After breath holding, pericardial rub continues but pleural rub stops.** Occasionally during inspiration, pericardial rub intensifies and this component is known as 'pleuro-pericardial rub'.

Case 78

SHAPE OF THE CHEST

Mention the common chest deformities :

1. Kyphoscoliosis.
2. Barrel-shaped chest.
3. Pigeon chest (pectus carinatum).
4. Funnel chest (pectus excavatum).
5. Alar chest (pterygoid chest).
6. Unilateral or bilateral, depression or fullness of the chest.
7. Deformity associated with ankylosing spondylitis, straight back syndrome, thoracoplasty operation (in this procedure, some ribs are resected on one side of chest to achieve collapse of the lung), severe chest trauma, or absence of part of chest wall (e.g., ribs, pectoralis muscle or clavicle).

Defects in the chest wall may be described in a better way like :

- Defects in sternum : Pectus carinatum, pectus excavatum.
- Defects in spine : Kyphosis, scoliosis, straight back syndrome, ankylosing spondylitis.
- Defects in costal cartilage : Rickety rosary, scorbutic rosary, costochondritis.

Kyphosis and scoliosis :

- (A) KYPHOSIS means backward bending of the vertebral column with its convexity posteriorly (like the shape of the letter 'K'). 'SCOLIOSIS' means lateral bending of the vertebral column (like the shape of the letter 'S').

These are abnormalities of alignment of the dorsal and lumbar spines. In severe kyphoscoliosis, they may lead to asymmetry of the chest wall resulting in restricted expansion of the chest wall, maldistribution of ventilation and blood flow in the lungs. Ultimately type II respiratory failure, pulmonary hypertension, RVF and chronic cor pulmonale develop.

- (B) Common causes are :

1. Congenital.
2. Vertebral diseases like ankylosing spondylitis, senile osteoporosis, osteoarthritis rheumatoid arthritis, rickets etc.
3. Caries spine— With 'gibbus' (acute angulation in the spine) formation.
4. Neurological causes— Neurofibromatosis, Friedreich's ataxia, syringomyelia, poliomyelitis and cerebral palsy.
5. Occupational, traumatic or postural.
6. Miscellaneous—Marfan's syndrome, Turner's syndrome, after thoracoplasty etc

Extensive unilateral fibrosis or collapse of the lung in childhood may give rise to scoliosis.

- (C) The common form of scoliosis with convexity to the right side displaces the apex beat towards the concavity, i.e., to the left side. Trachea may be displaced in simple scoliosis. It should be remembered that *The commonest cause of displacement of the apex beat is deformity of thoracic cage, usually scoliosis.*

(D) Method of diagnosis :

1. Scoliosis—The patient will stand erect with fully exposed chest and the observer looks for scoliosis from his back. It is observed whether the convexity is present in left or right side. Afterwards, it may be corroborated by palpation of the spine.
2. Kyphosis—The observer inspects the back from the sides in profile, i.e., a tangential view from both sides are necessary. The patient will stand erect with fully exposed chest.

In kyphosis, there is increase in the antero-posterior diameter of the chest.

* [LORDOSIS means forward bending of the vertebral column with its convexity anteriorly. It is commonly seen in pregnancy, obesity, secondary to hip joint disorders, muscular dystrophy, in large abdominal tumours or massive ascites].

Describe the normal chest :

1. Elliptical in cross section, i.e., transverse : antero-posterior diameter = 7:5
2. Bilaterally symmetrical and without undue elevation or depression.
3. Both the sides of the chest move simultaneously and symmetrically.
4. Subcostal angle is 90° or less than 90° (males having a narrower angle than females).

Barrel-shaped chest :

1. Antero-posterior diameter of the chest is increased (AP : TR=1 :1) and it is circular in cross section.
 2. The whole chest is relatively fixed in full inspiration.
 3. Intercostal spaces are full, ribs are less obliquely set and wide apart.
 4. There may be kyphosis of thoracic vertebrae.
 5. Fullness of supraclavicular fossae.
 6. The chest tends to move upwards in one piece.
 7. Wide subcostal angle.
 8. Prominence of sternal angle.
 9. Rarely encircling prominent veins are seen in the lower part of the chest.
- Cause : Severe obstructive airway disease or emphysema.

Pigeon chest (pectus carinatum) :

1. Triangular chest (in cross section) with prominent sternum.
 2. Rickety rosary (beaded appearance of the costochondral junction; best demonstrated in the 4th, 5th and 6th ribs).
 3. Harrison's sulcus (linear depression or sulcus present transversely from the sides of the xiphoid process to the midaxillary line as a result of repeated strong contractions of diaphragm).
- Causes :
- a) Childhood bronchial asthma.
 - c) Rickets.
 - b) Repeated lung infections since childhood.
 - d) Idiopathic.

Funnel chest (pectus excavatum) :

1. Curving backwards (i.e., depression) of the body of the sternum, usually the lower part.
2. Antero-posterior diameter < transverse diameter.
3. Undue prominence of the costochondral junction.

Causes :

- a) Repeated lung infections since childhood.
- b) In cobblers (also known as cobbler's chest).

* Pectus excavatum may give rise to displaced apex beat to the left and a 'benign systolic murmur due to right ventricular outflow tract obstruction; and the cardiac shadow may be enlarged in X-ray chest (PA view) as a result of sternal compression (Pomfret's heart). There may be restrictive type lung function defect, if very severe.

Alar chest :

1. Drooping of both the shoulders.
2. Winging of both the scapulae (i.e., prominent vertebral borders of scapulae).
3. Chest is narrow and tubular.

Cause : It is found in tall and thin built normal persons; often known as phthoid chest.

* Shield-like chest Broad chest with widely placed nipples, and is found in Turner's syndrome.

** Flail chest—Found in multiple rib fractures where the affected area moves inwards on inspiration, and outwards on expiration (paradoxical movement of chest).

Cross sectional view in different types of chest deformities :

1. Normal— Elliptical.
2. Barrel-shaped chest— Circular.
3. Pigeon chest — Triangular.

D/D of rickety rosary :

Scorbutic rosary (seen in scurvy)— The costochondral junctions are prominent, painful and appear sharp as well as angular; due to posterior subluxation or pushing in of the sternum.

Rickety rosary—The bead-like prominence at costochondral junctions are dome-shaped and semi-circular.

Fullness or depression of the chest :

(A) Fullness—

- | | |
|--|---|
| 1. Massive pleural effusion. | 7. Emphysema produces bilateral fullness. |
| 2. Empyema thoracis. | 8. Empyema necessitatis, aortic aneurysm, |
| 3. Pneumothorax. | or encysted pleural effusion produces |
| 4. Hydropneumothorax or pyopneumothorax. | 'localised' fullness. |
| 5. Bronchogenic carcinoma. | 9. Parietal oedema (e.g., from amoebic |
| 6. Pericardial effusion. | liver abscess). |

(B) Depression—

- | | |
|--------------------------|---|
| 1. Fibrosis of the lung | 3. Thickened pleura or pleural adhesions. |
| 2. Collapse of the lung. | 4. After rib resection in thoracotomy. |

Straight back syndrome :

The back becomes straight due to loss of normal thoracic kyphosis and thus, the antero-posterior diameter of the chest is reduced. The heart may be compressed between the sternum and the spine.

'Inspection' for shape and movement of the chest :

(A) Inspection of the chest is best done in standing or sitting (if standing is not possible) position, in good daylight with proper exposure (by stripping) of front and back of the chest. The patient should stand absolutely straight. Sitting means the patient will sit on a stool. The chest is inspected from:

1. Front.
2. Back.
3. The sides in profile.
4. Looking from above (standing behind the patient), over the shoulders for the upper part of the

If standing or sitting is not possible for the patient, then inspect the chest in lying down position (patient lies absolutely straight in the bed in supine position) from the :

1. Top (panoramic view).
2. Foot end of the bed.
3. The sides in profile.
4. Head end of the bed.
5. Back (try to turn the patient to any one side).

(B) Points to note :

1. Any deformity, fullness or depression (i.e., shape of the chest), apical impulse etc.
2. Back (winging of the scapula, drooping of the shoulder, kyphoscoliosis, gibbus, skin changes).
3. Whether both the sides of the chest are moving simultaneously and symmetrically.

** R Cad the Scheme of 'examination' of respiratory system under the heading 'Inspection'.

Classical winged scapula is found in paralysis of nerve to serratus anterior (C₅), sometimes in facio-scapulo-humeral muscular dystrophy, and severe malnutrition.

From the sides in profile and chest deformities (e.g., barrel-shaped chest) are best observed from the sides in profile.

In general survey, how do you examine 'Respiration' ?

Respiration should always be described by these 5 points.

- (A) Rate- Normal respiratory rate in a relaxed adult is 14-18 breaths/min though a wide variation occurs in health; at birth the rate is about 40/min. The variations are :
- Increased rate—Tachypnoea (read the section on 'Bronchial asthma').
 - Decreased rate—Bradypnoea (read the section on 'Bronchial asthma').
- * RATE OF RESPIRATION is counted surreptitiously (without the knowledge of the patient), by placing the hand in the epigastrium or with the fingers held on the radial pulse to avoid patients attention to breathing while observing the chest movements. The rate is counted for a full minute.
- (B) Rhythm—Changes in rhythm are diagnosed by inspecting the movement of the chest. There may be wide variations in health.
- Irregularly irregular or Blot's breathing—The respiration is sometimes slow, sometimes rapid, sometimes superficial, sometimes deep but without any constant relation of succession between the two types, with pauses at irregular intervals. Actually, there are 3-4 respirations without waxing and waning, followed by a pause. This is most commonly seen in meningitis, specially in children.
 - Regularly irregular or Cheyne-Stokes respiration (read the section on 'Bronchial asthma').
 - Miscellaneous—Kussmaul's breathing, stertorous breathing, prolonged inspiration or prolonged expiration (obstruction of upper or lower respiratory tract respectively).
- (C) Type—Diagnosed by inspecting the movement of the chest and abdomen. The variations are :
- Thoracic—Adult women, anxiety, peritonitis, huge ascites, hysteria, diaphragmatic palsy.
 - Abdominal—Adult men, young children, pleurisy, ankylosing spondylitis, intercostal muscle paralysis.
 - Abdomino-thoracic—Young children, sometimes in adult men.
 - Paradoxical respiration—Diaphragmatic palsy.
- * In females, intercostal muscles play more (so, thoracic type) and in males, diaphragm plays more (so, abdominal type) in normal respiration.
Females with predominantly abdominal type respiration—any painful condition in chest (e.g., pleurisy, chest trauma), pneumothorax etc.
Males with predominantly thoracic type respiration—any painful condition in abdomen (e.g., acute peritonitis), huge ascites etc.
- (D) Depth—Inspect the excursion of chest. The variations are :
- Shallow—Narcotic poisoning.
 - Deep—Hyperpnoea, hyperventilation e.g., in metabolic acidosis.
- (E) Breathing pattern—Wheeze or stridor, shallow breathing, air hunger, sighing, gasping, mouth-breathing or purse-lip respiration.
- * While examining the 'Respiration', one may comment on movement of accessory muscles of respiration or intercostal suction, if present.

Case 79**AUSCULTATION OF THE CHEST*****What will you do if asked to auscultate the chest ?***

If anyone is asked to auscultate the chest, he/she has to auscultate for both the respiratory system and CVS. During auscultation of left side of the chest, one has to be careful for both the systems and during auscultation of right side of the chest, only respiratory system will serve the purpose. Auscultate over front (inflacavicular area to lower costal margin), sides (axillary and in ra axillary areas) and back (supra-, inter- and infrascapular areas) avoiding auscultation over bones (clavicle, sternum and scapula). Always compare with the other side.

If a particular area of the chest (suppose, the area below the right nipple) is asked to be auscultated one must take the permission of the examiner for auscultation over the identical site on left side of the chest for observations regarding vocal resonance (always say vocal resonance as normal increased or decreased after comparing with the other side).

(A) **For respiratory system**, the description will be in the order of :

1. Breath sounds,
2. Vocal resonance, and
3. Adventitious sounds.

(B) **For CVS**, the description goes like this :

1. Heart sounds S₁, S₂, any S₃ or S₄ (quality, intensity, splitting etc.),
2. Murmur,
3. Pericardial rub, ejection click, opening snap, pericardial knock etc.

* In the respiratory system, auscultation over trachea is not at all necessary.

During auscultation of the chest, direction given to the patient :

The clinician will stand on the right side of the patient. Before auscultation, the patient is asked to :

- a) Turn the face to the left side (so that patient can not breathe on the clinician's face),
- b) Breathe through open mouth (to prevent noise from partially closed nose),
- c) Breathe regularly and deeply, but without any noise.

* Demonstrate the open mouth respiration to the patient for easy understanding.

How to classify the breath sounds ?

Breath sounds- Sounds produced by the passage of air through the tracheo-bronchial tree upto the alveoli. Gas turbulence in the major airways is responsible for inspiratory sound and elastic recoil of the lung for expiratory sound. The breath sounds are divided into three types (mainly two types : vesicular and bronchial) :

(A) Vesicular breath sound—

- a) Definition : It is the sound produced by the passage of air in and out of the alveoli (vesicles)
- b) Characteristics :
 - (i) Rustling (sound like dry leaves blown by the wind) in character.
 - (ii) Intensity of inspiration is more than that of expiration.
 - (iii) Duration of inspiration is greater than that of expiration.
 - (iv) No gap in between inspiration and expiration.

* As the normal lung tissue selectively filters out some higher frequencies, the breath sound becomes quieter, and results in vesicular breath sound (the normal breath sound over the chest).

c) Classical sites :

Inframammary, infra-axillary and infrascapular regions.

d) Common variations :

1. Diminished vesicular— Thick chest wall, obesity, thickened pleura, pleural effusion, ^ pneumothorax, hydropneumothorax, empyema thoracis, fibrosis, emphysema etc.
2. Vesicular breath sound with prolonged expiration— Bronchial asthma, chronic bronchitis, emphysema. In this type of respiration, the duration of inspiration and expiration becomes more or less equal.
3. Harsh vesicular—
 - (i) Intensities of inspiration and expiration are increased.
 - (ii) Usually pathological.
 - (iii) Found in compensatory emphysema, forced breathing.
4. Puerile—
 - (i) Intensities of inspiration and expiration are increased.
 - (ii) Normal physiological phenomenon.
 - (iii) In thin built persons (specially children).
5. Cogwheel or jerky— Patients who are hysterical, very nervous or crying during auscultation,
- b. Absent breath sound— Pneumothorax, massive pleural effusion, hydropneumothorax collapse with obstructed bronchus, grossly thickened pleura etc.

The quality of breathing in harsh and puerile types remain the same.

(B) Bronchial breath sound—

- (a) Bronchial ^{T¹ produced by the} Passage of air through the tracheo-bronchial tree. ^{I r n n u i s o u ? 15 audible in} situations where sound generated in central airways is pathology (e.g., consolidation) with a patent bronchus. ^{^ H H S S H U n ? T d U l r O U 8 h t h C l u n g s u b s t a n c e s h a v i n g u n d e r m e n t i o n e d}

(b) Characteristics :

- (i) 'Blowing', hollow or 'aspirate' in character.

- (ii) Intensity of expiration is more than that of inspiration.
- (iii) Duration of expiration is usually equal to inspiration.
- (iv) Definite gap present in between inspiration and expiration.
- (c) Classical site : Over the trachea.
- (d) Types :
 1. Tubular— This is a high-pitched bronchial breath sound. It is commonly found in,
 - (i) Consolidation.
 - (ii) Collapse of the lung with patent bronchus.
 - (iii) Sometimes, above the level of pleural effusion.
 - (iv) Many a time, tubular breath sound is heard if the trachea or large bronchus physically comes near the stethoscope by push-pull effect (e.g., fibrosis of the lung)—loosely termed as 'pulled tracheal syndrome'.
 2. Cavernous— It is a low-pitched bronchial breath sound classically heard over a superficial, big, empty cavity connected with a patent bronchus e.g., tuberculous cavity, lung abscess.
 3. Amphoric—It is a low-pitched bronchial breath sound with tones and overtones, or with a metallic tone. The sound mimics the whistling sound produced by blowing air across the mouth of a small glass bottle. This is classically seen in,
 - (i) Bronchopleural fistula (open pneumothorax), and
 - (ii) Occasionally in a big cavity connected with a small bronchus.

* **Conditions associated with bronchial breath sound will produce quantitative increase in vocal resonance, i.e., bronchophony and whispering pectoriloquy.**

** In obstructed main bronchus, bronchial breath sound may not be heard over consolidation or collapse.

(C) Bronchovesicular breath sound (going to be obsolete though few old school clinicians still prefer it)—

- a) Definition :
This is a mixture of two breath sounds where the expiration is mainly of bronchial and inspiration is predominantly of vesicular in character, and there is no gap in between inspiration and expiration.
- (b) Classical sites (in health) :
 1. Right infraclavicular area (here, trachea is in direct contact with the lung whereas in the left infraclavicular area the aorta, oesophagus and internal carotid artery separate the trachea from the lung).
 2. Interscapular area at the back (root of the lung).
- (c) Clinical associations :
 1. Consolidation in the stage of resolution.
 2. Partial collapse of the lung.
 3. Cavity in the lung.

Caution during auscultation of respiratory system :

1. *Always try to auscultate with the diaphragm of stethoscope* to avoid time consumption and practical difficulty; as most of the normal lung sounds are low-pitched, the bell is preferred over the diaphragm by some clinicians.
2. Ask the patient to cough :
 - a) To clear the throat (if throat sounds appear).
 - b) To note the changes in crepitations (often helps in differentiation with pleural rub).
 - c) For auscultating post-tussive crepitations (specially in tuberculous cavity).
3. Press the diaphragm of stethoscope in patients with pleural rub to note the local tenderness and enhancement of the intensity of pleural rub.

What is Valsalva manoeuvre ?

This is an attempted **forced expiration** in closed glottis when the mouth is shut and the nose is held closed. This manoeuvre increases the intrathoracic pressure and thus obstructs the venous return in the right atrium. Classical example is the straining exerted during defecation.

Clinical applications ;

1. To make the abdominal wall veins prominent with the help of Valsalva manoeuvre while the patient sits in his bed.

2. During the Valsalva manoeuvre—Tachycardia, overshoot of BP and bradycardia occur sequentially in health. This phenomenon is found to be absent in CCF, ASD and autonomic neuropathy.
3. Murmur of mitral valve prolapse syndrome (MVPS) and HOCM increases during Valsalva manoeuvre.
4. As a vagotonic procedure to abolish ectopic beats.
5. To terminate bouts of hiccough as a simple household remedy.

What is Muller's manoeuvre ?

It is just the opposite of the Valsalva manoeuvre. This manoeuvre is an attempted forced inspiration in closed glottis when the mouth is shut and the nose is held closed. This manoeuvre decreases the intrathoracic pressure. Classical example is a drowning man.

Case 80

PLEURAL RUB

Characteristics :

It is a leathery or creaking sound produced by the rubbing of inflamed and roughened, visceral and parietal pleura. The pleural rub is,

1. Superficial (the sound seems to be very close to the ear), scratchy or grating in quality.
2. Biphasic (heard during inspiration and expiration both).
3. Better heard on pressing the chest piece of stethoscope over the chest wall.
4. Localised—Generally heard over the antero-inferior part of lateral chest wall or in the lower part of back as the movement of lung is maximum in these regions.
5. Neither any alteration after coughing nor any variation with change of posture,
6. Patient may complain of pain in the chest and local tenderness may be elicited.
7. Disappears when breath is held.
8. At times it is palpable—The 'friction fremitus'.

Differential diagnosis :

- (A) It should be differentiated from coarse crepitations (which is also biphasic) by the following points. Crepitations are :
 - (i) Not intensified on pressing the stethoscope,
 - (ii) Modified or diminished after coughing,
 - (iii) May be auscultated over a widespread area, and
 - (iv) Discontinuous; not superficial, not associated with pain or tenderness.
- (B) Pericardial rub (pericardial rub continues after holding the breath).
- (C) Hepatic or splenic rub—often very difficult to differentiate, and is audible over enlarged liver and spleen respectively. Hepatic and splenic rub are inaudible if the patient holds his breath.

* Occasionally, after pressing the stethoscope in an uneven chest (with prominent ribs), pleural rub-like sound is produced due to friction of moist, sweaty chest wall with the diaphragm of stethoscope. Put the chest piece in a new place near the previous one after wiping off the sweat.

** In a hairy chest wall, avoid friction of hair by moistening the chest wall with sprinkled water and henceforth apply the chest piece tightly over the chest.

What is your diagnosis ?

Acute dry pleurisy.

Causes of acute dry pleurisy :

- | | |
|---|-----------------------------------|
| 1. Pulmonary tuberculosis. | 7. Collagen vascular diseases. |
| 2. Pneumonic consolidation. | 8. Subdiaphragmatic abscess. |
| 3. Bronchogenic carcinoma. | 9. Bronchiectasis. |
| 4. Pulmonary infarction. | 10. Injury to the chest wall. |
| 5. Bornholm disease (epidemic pleurodynia). | 11. Pleural metastases. |
| 6. Amoebic liver abscess. | 12. Mesothelioma of pleura (rare) |
- 1, 2, 3 and 4 are most common and important causes.

Classical chest pain of acute dry pleurisy (pleuritic pain) :

1. Sharp, stabbing or tearing localised pain.
 2. It increases on coughing, sneezing, deep inspiration, pressure from outside, jolting or bending.
3. Localised pain (usually non-radiating).
4. The pain is usually non-exertional but may increase after exercise (due to tachypnoea).
5. When the diaphragmatic pleura is involved, the pain may be referred to the tip of shoulder (central part of the diaphragm is supplied by C₃₄₅) or to the epigastrium (domes of the diaphragm are supplied by intercostal nerves).
6. The pain is diminished by holding the chest or holding the respiration (i.e., by taking shallow respiration).
7. Associated with pleural rub.

* Pleuritic chest pain is often associated with pyrexia and dry cough. See also page 69.

** Parietal pleura is pain sensitive (supplied by intercostal and phrenic nerve), while visceral pleura is pain insensitive (supplied by autonomic nervous system).

Decubitus in different diseases of pleura :

- (A) Pneumothorax— Patient prefers to lie on the affected side. It allows maximum expansion of the normal lung.
- (B) Pleural effusion— Patient prefers to lie on the side of effusion as it allows maximum expansion of the unaffected lung. Pleural fluid may compress the mediastinum and produce discomfort, if the patient lies on the normal side.
- (C) Acute dry pleurisy—
 1. Majority of patients lie on the affected side (if there is absence of overlying sensitive skin). In this way they splint their chest wall with the surface of the bed.
 2. Few patients lie on the opposite side because the overlying skin is very sensitive and the patient cannot keep contact with the bed.

Describe different types of 'decubitus' in clinical medicine :

Decubitus means posture or attitude of the patient in bed assumed by him/her in most of the time of day and night. The different variations are :

1. Of choice—The patient is comfortable in any position in bed (also known as 'dorsal decubitus' or 'supine' position).
2. Propped-up— Propped-up position with a back-rest is found in patients suffering from dyspnoea. The back-rest can be adjusted according to the requirement of the patient e.g.,
 - Mild dyspnoea— adopts 45°
 - Moderate dyspnoea— adopts 60°
 - Severe dyspnoea or orthopnoea— adopts 90° (i.e., patient sits in bed).

Dyspnoea is basically due to respiratory and cardiovascular ailments (cardio-respiratory embarrassment). Gastrointestinal cause like massive ascites, and neurological causes like diaphragmatic palsy, respiratory muscles palsy, myasthenia gravis may also produce dyspnoea. Propped-up decubitus relieves dyspnoea because,

- a) The excursion of diaphragm occurs better in erect posture.
- b) Erect posture diminishes the venous return and thus dyspnoea of cardiac origin becomes less.
- c) While in recumbency, the increase in intrathoracic blood volume elevates pulmonary venous and capillary pressure which ultimately increase the pulmonary closing volume and diminishes vital capacity. This is corrected on assuming erect posture.
3. Stooping forward position (Mohammedan's prayer position) on a cardiac table— Seen in severe dyspnoea of pericardial effusion, acute severe asthma, cardiac asthma and COPD. It is also observed in acute pericarditis or acute pancreatitis to get relief of pain. Patients with acute pancreatitis is partly relieved by lying down with knees drawn towards the chest.
4. Squatting—Fallot's tetralogy is the commonest cyanotic congenital heart disease where squatting is observed. As soon as the patient becomes dyspnoeic, he adopts squatting posture to relieve from dyspnoea. Squatting increases the peripheral resistance (i.e., pressure in aorta increases) and thus reduces the right-to-left shunting through VSD.
5. Trepopnoea—See page 98.
6. Platypnoea—See page 98.

7. Lateral decubitus—See above (decubitus adopted in diseases of pleura); patient of liver abscess prefers to lie on the left side.
8. Listless attitude or where the patient lies still in bed. This attitude is usually seen in peritonitis (abdominal wall movement causes intense pain), anginal pain, coma and hysteria.
9. Curled-up—Observed in renal colic and biliary colic; restless, tossing—In colics, AMI.
10. Head end down (usually adjusted by placing bricks or wooden blocks in the foot end of bed)—In the treatment of bulbar palsy, after lumbar puncture, after spinal anaesthesia, balanced traction in orthopedics, in treatment of sinus bradycardia developed from acute myocardial infarction, deep venous thrombosis in legs, and in postural drainage of bronchiectasis and lung abscess (lower lobes).
11. Decubitus of hemiplegia—The affected arm remains flexed, adducted and semipronated, and the affected lower limb adopts extended, adducted and plantiflexed attitude. As a whole, the affected side shows less movement while the patient is in bed.
12. Opisthotonus, pleurothotonus and emprosthotonus—Respectively, they are bending of the body like an arc in forward direction, laterally to one side, and backward direction. These bending postures are seen in tetanus and strychnine poisoning.
13. Dorsal decubitus position with a pillow placed below the legs (pedal oedema, cellulitis of foot) or below the popliteal fossa (acute inflammation of knee joint, e.g., in rheumatic and rheumatoid arthritis, haemophilia).
14. Decerebrate posture—Extended elbows and wrists, with arms pronated; there is tonic extension and plantiflexion of the lower extremity, associated with head retraction and jaw clenching. The lesion lies at upper brainstem level, disconnecting cerebral hemispheres from brainstem.
15. Decorticate posture Flexed elbows and wrists, with arms supinated; lower extremities show tonic extension. It is seen in bilateral hemispherical lesion above midbrain.
16. Prone position—Colics, abdominal aortic aneurysm (eroding vertebra and producing back pain).
17. Knee-elbow position—Pain of chronic pancreatitis is reduced on assuming this posture.

Medical (non-surgical) causes of pain abdomen :

1. Basal pneumonia (basal pleurisy).
2. Acute myocardial infarction (inferior wall infarction commonly).
3. Diabetic ketoacidosis (may be due to acute gastric dilatation and/or pancreatitis).
4. Henoch-Schonlein purpura (vasculitis).
5. Sick cell anaemia (vaso-occlusive crisis).
6. Acute intermittent porphyria (autonomic neuropathy).
7. Intercostal herpes zoster (pre-herpetic eruption produces radicular pain).
8. Caries spine or cord compression (radicular pain); collapse of vertebra.
9. Tabetic crisis (probably due to autonomic neuropathy; not seen now-a-days).
10. Polyarteritis nodosa (vasculitis).
11. Allergic pain (C_t esterase inhibitor deficiency).
12. Hyperlipidaemia (familial).
13. Lead poisoning (colic).
14. Localised tetanus of anterior abdominal wall muscle.
15. Functional gastrointestinal disorders.

* Peptic ulcer, acute pyelonephritis and acute pancreatitis may also be considered.

** Endocrine causes of pain abdomen are : (i) Diabetic ketoacidosis, (ii) Adrenal crisis, (iii) Hyperparathyroidism, and (iv) Cushing's syndrome.

Pain abdomen associated with shock :

- | | |
|--|-------------------------------------|
| 1. Acute pancreatitis. | 7. Rupture of amoebic liver abscess |
| 2. Peptic ulcer complicated by bleeding or perforation. | 8. Adrenal crisis. |
| 3. Acute myocardial infarction (inferior wall infarction). | 9. Acute intermittent porphyria. |
| 4. Diabetic ketoacidosis. | 10. Strangulation of hernia. |
| 5. Acute mesenteric ischaemia. | 11. Ruptured ectopic pregnancy. |
| 6. Dissection of abdominal aorta. | 12. Septic abortion. |

Absent pleural rub after few days—reasoning behind :

Either the patient is cured, or developed pleural effusion.



Exfoliative dermatitis (erythroderma) from internal malignancy (lymphoma)



Extensive skin lesions in **Stevens-Johnson syndrome** with signs of healing



Multiple **erythema nodosum** (panniculitis) in sarcoidosis



Erythema multiforme with bullous (Stevens-Johnson syndrome) and iris / target (red periphery with a cyanotic hue) lesions



Painless **oral (palatal) ulcer** in systemic lupus erythematosus



Pityriasis versicolor – manifested by hypopigmented macules in right side of the face



Viperidae snake bite with cellulitis, oedema and haemorrhagic blister formation in and around the site of bite



Salt-pepper appearance in skin [depigmentation (white-like salt) and pigmentation (black-like pepper)] in **scleroderma**



Acanthosis nigricans – brown to black, poorly defined, velvety hyperpigmentation of the skin of neck in type 2 diabetes mellitus (indicating insulin resistance) with skin tags (acrochordons)



Elephantiasis of legs from **filariasis**

Management :

1. Rest in bed.
2. Relief of chest pain by,
 - a) Hot fomentation.
 - b) Splinting of the chest wall with leukoplast.
 - c) Analgesics (NSAIDs).
 - d) Sedatives.
3. Treatment of the underlying aetiology.

Identification points :

1. Sometimes, it may be confused with pericardial rub (in left-sided pleurisy). Pleural rub stops after holding the breath but pericardial rub continues.
2. It should be differentiated from coarse crepitations by the points discussed above.

Conclusion :

To diagnose pleural rub, remember these 3 points :

1. Hold the breath (pleural rub disappears while pericardial rub continues).
2. Ask the patient to cough (D/D with crepitations; pleural rub does not alter with cough).
3. Press the chest piece of stethoscope (pleural rub intensifies or patient may C/O pain).

Case 81

EXAMINATION OF THE TONGUE

How examination of the tongue helps in the diagnosis in clinical practice ?

- (A) Ask the patient to SHOW THE TONGUE by protruding it outside the oral cavity—the patient is unable to protrude the tongue in case of short frenum (known as 'tongue tie' or ankyloglossia), malignancy of the tongue and also in bilateral paralysis of the tongue (XIIth nerve palsy). Sluggish and slow protrusion is seen in mental retardation.
- (B) MOVEMENT OF THE TONGUE :
 1. **'Trombone tongue'** — rapid forward and backward movement of the tongue is characteristic of general paralysis of insane (GP1); may be seen in Parkinson's disease.
 2. Ask the patient to protrude the tongue — the patient protrudes it momentarily and takes it back within the oral cavity instantaneously (lizard tongue)—seen in rheumatic chorea (Jack-in-the-box tongue).
 3. 'Chewing tongue' — seen in athetosis, as if the patient is chewing something.
 4. Irregular and continual rotatory movements of the tongue may be seen in extrapyramidal syndromes (dyskinesia) induced by drugs like phenothiazines, metoclopramide, levodopa.
 5. Rolling movements of tongue are found in cretins, mongols and as part of tics.
 6. Ask the patient to protrude the tongue out of the oral cavity and look for fine **tremor**—characteristically found in anxiety neurosis, thyrotoxicosis, chronic alcoholism etc.
 7. Ask the patient to keep the tongue relaxed in the floor of the mouth and look for **fasciculation**; it is seen in MND, bulbar palsy or LMN palsy of XIIth cranial nerve.
 8. Observe for lingual myoclonus (with palatal myoclonus), forced deviation of tongue (as part of focal seizure), and tics and habit spasm.
- (C) SIZE OF THE TONGUE :
 1. Large tongue or **macroglossia** (with impression of teeth in sides) is seen in,
 - (i) Acromegaly.
 - (ii) Myxoedema.
 - (iii) Cretinism.
 - (iv) Down's syndrome.
 - (v) Primary amyloidosis.
 - (vi) Mucopolysaccharidosis e.g., Hurler syndrome.
 - (vii) Haemangioma or lymphangioma of tongue.
 - (viii) von Gierke's disease (glycogen storage disease).
 - (ix) Angioneurotic oedema.
 2. Small tongue or **microglossia** is seen in,
 - (i) Cerebral diplegia.
 - (ii) Bulbar and pseudobulbar palsy.
 - (iii) Motor neurone disease (MND).
 - (iv) Wasting of the tongue (LMN lesion of XIIth cranial nerve).

- * Acute swelling of tongue is found in bite, infection, angioneurotic oedema, haemophilia, pemphigus.

(D) COLOUR AND MORPHOLOGICAL CHANGES :

1. Dry tongue — dehydration, mouth breathing, xerostomia, after administration of atropine.
2. Moist tongue — sialorrhoea in post-encephalitic parkinsonism, heavy metal poisoning.
3. Pale tongue — anaemia (severe).
4. Blue tongue — central cyanosis, meth- or sulphaemoglobinaemia (mainly the sides of the tongue), intake of blue coloured food material.
5. Yellow tongue — jaundice (the undersurface), intake of yellow coloured sweets.
6. Bluish-red tongue — polycythemia.
7. **Magenta-coloured tongue** (a bit pinkish) — riboflavin deficiency.
8. Black tongue — after ingestion of bismuth, liquorice, charcoal; Addison's disease.
9. Blotting paper like pallor (with black pigmentation in the margin) — often seen in hook-worm infestation.
10. White or greyish coating or 'furred' tongue — smoking, chronic debility, acute tonsillitis, sore throat etc. 'Fur' is common in heavy smokers.
11. **'Bald tongue'** — there is total loss or atrophy of papillae, and is classically seen in pellagra, pernicious anaemia, iron deficiency anaemia, tropical sprue and syphilis.
12. **'Raw-beefy tongue'** — red, swollen and painful tongue. Found in pellagra and vitamin B₁₂ deficiency.
13. **'Angry-looking' tongue** — central coating with red tip and margins (enteric fever).
14. 'Black hairy' tongue — produced due to failure of keratin layer of the filiform papillae to desquamate normally. It is found in staining by tobacco, food, chromogenic organism, fungal infection, after use of penicillins and tetracyclines.
15. 'Geographic' tongue — denuded red patches 'wandering' or migrating (look like boundaries of a country map) across the surface of the tongue due to rapid loss and regrowth of filiform papillae. It is an asymptomatic inflammatory condition and though looks odd, it has no clinical significance.
16. 'Strawberry' and 'raspberry' tongue — hypertrophy of fungiform papillae with changes in filiform papillae, and is seen in scarlet fever. The tongue looks red with papillae standing out as white dots.
17. 'Scrotal' tongue — deep horizontal fissures in the tongue where debris may collect; of no clinical significance.
18. Median rhomboid glossitis — a lozenge-shaped denuded area in the middle of the tongue posteriorly. It is a congenital abnormality and should be differentiated from carcinoma of the tongue as it feels nodular.
19. White patches on the tongue — due to thrush (i.e., moniliasis due to immunosuppressive therapy, diabetes and AIDS), other fungal infection, leucoplakia, chronic superficial glossitis etc.
20. Ulcers in the tongue — aphthous ulcer, frenal ulcer (in frenum), tuberculous ulcer (in dorsum), malignant ulcer (anywhere in the tongue), snail-track ulcer (in dorsum) in secondary syphilis, ulcers at the margins (by ill-fitted dentures) or ulcers from Behcet's disease (mimicking aphthous ulcer) may be present.
21. Bite mark in the tongue — accidental bite during eating or after convulsions.
22. **Spastic tongue** with pointed tip (without any fasciculation) — in pseudobulbar palsy.
23. **Flaccid tongue** with rounded tip and the tongue sits on the floor of the mouth like a mushroom (grossly wasted), often with fasciculation — in bulbar palsy.
24. Growth in the tongue (with halitosis)* — in squamous cell carcinoma of the tongue.
25. Mushroom like tongue — sore tongue with white slough, seen in corrosive poisoning.
26. Horny tongue (crocodile tongue) — tongue with cornification of the mucosa.
27. Dry, red tongue with atrophy of the papillae and fissures (parchment-like tongue)—seen in Sjogren's syndrome.
28. Hairy leucoplakia—painless white corrugated lesion on sides are due to Epstein-Barr virus infection, specially in AIDS.

(E) DEVIATION OF THE TONGUE :

Ask the patient to protrude his tongue,

1. In UMN lesion of XIIth cranial nerve — deviated to the opposite side.
2. In LMN lesion of XIIth cranial nerve — deviated to the same side.

(F) POWER OF THE TONGUE ;

Ask the patient to move the tongue inside the oral cavity at random and to press the cheeks with the tongue when resistance is applied in the cheeks from outside.

(G) TASTE SENSATION :

Taste sensation of anterior 2/3rd of the tongue is carried by the facial nerve and that of posterior 1/3rd is carried by the glossopharyngeal nerve. Sugar or saccharin, common salt, vinegar and quinine are used to test the sweet, salt, sour and bitter taste sensations respectively. Sensations perceived by the tongue are sweet at the tip, sour at the margins, bitter at the back and salt by any part of the tongue. Loss of taste sensation is ageusia and may result from lesions in any part of the peripheral and central pathways of taste fibres.

Directly ask the patient for recent loss of taste or any abnormal taste sensation. Now to test the taste sensation, ask the patient to close his eyes, hold the tongue with a swab and put the substance on the anterior 2/3rd of the protruded tongue on each side in turn. Ask him to identify the substance. Each time mouth is washed and a new substance is applied. Bitter sensation is tested last.

* **Halitosis** (malodorous breath)—the common causes are bad oral hygiene, smoking, decomposed food debris collected in between teeth, stomatitis, gingivitis, caries tooth, sinusitis, atrophic rhinitis, any tumour of nasal passage, carcinoma of the tongue, bronchiectasis, lung abscess, diabetes, hepatic and renal failure, gastro-colic fistula and intestinal obstruction.

** **Sialorrhoea** or ptyalism (hypersalivation) occurs during pregnancy, carcinoma of the tongue or mouth, stomatitis, caries tooth, Wilson's disease (drooling), acid or alkali poisoning, post-encephalitic parkinsonism, bulbar palsy, schizophrenia, hydrophobia; in arsenic, mercury or lead poisoning.

*** **Xerostomia** (dry mouth) is common in fear, anxiety, dehydration, high fever, drug-induced (diuretics, antipsychotics), Sjogren's syndrome, as a complication of polyuria, irradiation for head and neck carcinoma and psychogenic.

**** pigmented lesions within the oral cavity are seen in Addison's disease, Peutz-Jegher s syndrome, haemochromatosis, melanoma and lead poisoning (blue line or Burtonian line in the gum).

***** Tongue may be regarded as mirror of dysfunction of all the systems; previously tongue was known as the mirror of G. I. tract only.

Case 82

VISIBLE PERISTALSIS

Significance :

Usually no peristaltic wave is seen in health. Visible peristalsis signifies mechanical obstruction of intestine (pathological). As a normal finding, it is occasionally seen in children with thin abdominal wall or in adults where the abdominal musculature has been thinned out by repeated pregnancy, long-continued ascites or in elderly patients with lax abdominal wall (physiological).

Localisation of obstruction :

Obstruction is generally seen in one of these three places like stomach, small gut or large gut, and the site of obstruction is determined by analysing the peristaltic waves.

- a) **STOMACH** — Usually visible peristalsis is seen due to obstruction at pylorus [Children - congenital hypertrophic pyloric stenosis (CHPS), adult - pyloric stenosis from chronic duodenal ulcer, old age - pyloric growth may produce gastric outlet obstruction] and the wave moves in the upper "abdomen from left to right hypochondrium, occasionally through the umbilical region. These waves are golf-ball like and generally not more than 2-3 waves are seen at a time. _
- b) **SMALL GUT** — The peristaltic waves are seen as 'ladder pattern' (zigzag fashion) in the peri-umbilical region and there may be 10-12 waves formation per minute,
- c) **LARGE GUT** — If the obstruction is located in transverse colon, the waves are seen moving from right to left hypochondrium (opposite to pyloric obstruction). These are larger in size, i.e., cricket-ball like. The waves are seen in the flanks in case of obstruction of ascending or descending colon.

How to evoke peristalsis at the bedside ?

In a suspected case, peristalsis can be Induced by,

1. Flicking or gentle massage of the anterior abdominal wall, or
2. Drinking water (1-2 glasses), or
3. Pouring few drops of ether or alcohol over the abdomen.

Regarding inspection of waves, a tangential view is always better for slow waves. Small intestinal peristalsis may be occasionally seen normally through divarication of recti or in incisional hernia through abdominal scar.

Common causes of gut obstruction :

- (A) Stomach - Pyloric stenosis, carcinoma of the stomach.
- (B) Small gut - Adhesions and external hernias.
- (C) Large gut - Colonic carcinoma, sigmoid diverticulitis and volvulus.

Common causes of gut obstruction in India :

Round worm infestation in children and intestinal tuberculosis in any age group.

Clinical confirmation, if peristalsis is seen in upper abdomen :

Give the patient a quick shake and carefully listen for succussion splash (e.g., pyloric stenosis).

Confirmation of diagnosis of intestinal obstruction :

After careful history taking and meticulous clinical examination, the physician should do a straight X-ray of the abdomen **in erect posture**, where observation of multiple fluid levels will confirm the diagnosis (normally there may be fluid levels in i) stomach ii) lower duodenal flexure, and at iii) ileo-caecal junction). *More than three fluid, levels clinch the diagnosis of intestinal obstruction.*

Importance of auscultation over abdomen :

1. Peristalsis Normal sound (vide page 111); increased in intestinal obstruction; absent in paralytic ileus.
2. Bruit—Specially in renal artery stenosis (best heard just above the umbilicus, about 2 cm on either side of midline), coarctation of aorta and aortoarteritis of abdominal aorta (systolic bruit); hepatic bruit (vide page 283). Bruit is also heard over aortic aneurysm.
3. Succussion splash—Vide page 110 (pyloric stenosis).
4. Venous hum—Vide page 434.
5. Hepatic and splenic rub—Vide page 283 and 287 respectively.
6. Ausculto-percussion—Vide page 110.
7. Uterine souffle (pregnancy).
8. Foetal heart sounds (auscultate with the bell of stethoscope).

Conclusion :

In a patient with visible peristalsis, always localise the 'site of obstruction' and look for :

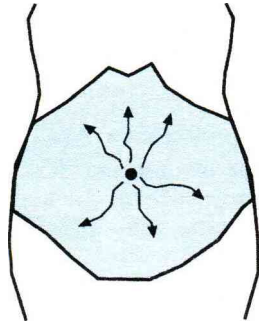
- 1) Incision mark in the abdomen (adhesion from past abdominal operation),
- 2) Succussion splash, and
- 3) Borborygmi (intestinal obstruction).

Case 83**PROMINENT VEINS WITH VENOUS HUM****What is venous hum :**

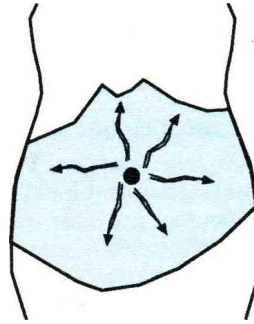
It is synonymous with murmur heard in arteries. One may call it venous murmur. It is produced as a result of enormous blood flow through the veins.

Common sites for venous hum :

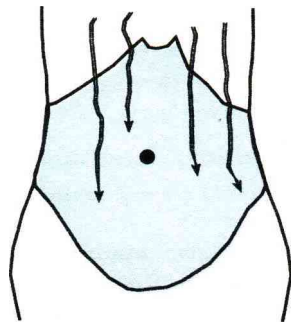
Commonly heard as a continuous humming sound over prominent veins (i.e., collaterals) seen in the epigastrium or dilated veins seen around the umbilicus in cirrhosis of liver (pathological). Some-

**Normal**

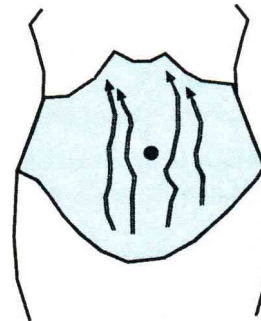
Direction of flow is away from the umbilicus
(no venous prominence is seen in health)

**Portal hypertension**

Direction of flow is away from the umbilicus
(both above and below the umbilicus)

**SVC obstruction**

Direction of flow is from 'above downwards'
whether it is above or below the umbilicus
(mainly the chest wall veins are seen)

**IVC obstruction**

Direction of flow is from 'below upwards',
whether it is above or below the umbilicus

Fig. 2 : Abdominal wall veins

times it is heard above either clavicle in a child and during pregnancy (physiological). The cervical venous hum (Devil's murmur) in adults (pathological) is often audible in hyperkinetic circulatory states e.g., severe anaemia, thyrotoxicosis etc.

Characteristics of venous hum :

1. Soft and low-pitched.
2. Often continuous with early diastolic accentuation.
3. Best heard in sitting or erect position with the bell of stethoscope.
4. Best audible in inspiration.
5. Disappears on pressing the bell of stethoscope (so, place the bell lightly over a prominent vein), and after Valsalva manoeuvre. It is accentuated by exercise.
6. Thrill may be associated with and detected by application of light pressure.

In which situation should we search for abdominal venous hum ?

In the presence of dilated abdominal veins, we search for venous hum. The combination of dilated abdominal wall veins (caput medusae) and a loud venous hum at the umbilicus with normal liver is called Cruveilhier-Baumgarten syndrome. This syndrome may originate due to congenital patency of umbilical vein but more usually to a well-compensated cirrhosis.

Direction of blood flow in these prominent abdominal veins (see question above) :

Blood flow is always away from the umbilicus as it is dealing with a case of 'portal hypertension'.

D/D of venous hum :

- (A) In the epigastrium : Hepatic bruit (over liver) or continuous murmur (e.g., coarctation of aorta).
- (B) In the upper chest or supraclavicular area (left side) : Patent ductus arteriosus.

How these abdominal wall veins are formed in portal hypertension ?

Dilated and tortuous veins suggest venous obstruction. These are formed due to blood passing through a large umbilical or paraumbilical vein in the falciform ligament **from** the left branch of portal vein **to** the superior epigastric, internal mammary or inferior epigastric veins present in the abdominal wall (i.e., opening of anastomotic channels between portal and systemic veins).

How these abdominal wall veins are formed in IVC obstruction ?

These are dilated anastomotic channels formed between the superficial epigastric and circumflex iliac vein **below**, and the lateral thoracic veins **above**, carrying blood from long saphenous vein to axillary vein. This is why the direction of blood flow is from below upwards.

Conditions associated with dilated chest wall veins

1. SVC obstruction (commonest).
2. Severe congestive cardiac failure.
3. IVC obstruction (not uncommon).
4. Localised venous prominence : underlying pleural malignancy or empyema thoracis.
5. Extreme cachexia (due to loss of subcutaneous fat), and sometimes in lean and thin persons.
6. Thrombophlebitis of anterior thoracic vein (Mondor's disease)—rare.

How to know the presence of dilated and tortuous abdominal / chest wall veins ?

1. The patient is asked to sit with legs hanging from the bed (never examine in lying down position).
2. Then the patient is asked to **cough** or to perform the **Valsalva manoeuvre**. Cough makes the veins prominent transiently while the Valsalva retains the prominence of veins so long the manoeuvre is continued.
3. Proper light is necessary (patient facing the window) for demonstration.

Detection of direction of venous blood flow :

Method :

1. Make the veins prominent by adopting the above mentioned method. A tributary-free, long segment of vein (preferably one inch or more) is selected for examination.
2. Stand on the right side of the patient and place the index finger of both hands side by side over the vein—the left above the right one. Now start milking the vein by movement of two index fingers in opposite direction.
3. The two ends of the bloodless vein is blocked with the pressure given by two index fingers. The left index finger is now removed and the rapidity of venous filling from above is noted. The same procedure is repeated after removal of the right index finger and the rapidity of venous filling from below is observed now. The rapidity of venous filling indicates the direction of blood flow.

Venous flow in different conditions (Fig. 2 in page 435) :

- (A) **Normally** — Away from the umbilicus (but one can not see any venous prominence in health).
- (B) **Portal hypertension** — Away from the umbilicus (both above and below the umbilicus).
- (C) **FVC obstruction** As a whole, the flow is below upwards, i.e.,
 - (i) Above the umbilicus - Upwards and away from the umbilicus.
 - (ii) Below the umbilicus - Towards the umbilicus.
- (D) **SVC obstruction** — From above downwards as a whole.

Diagnosis of these pathological conditions at bedside :

The classical direction of venous flow helps in the diagnosis and the added features are •

1. Portal hypertension - Pedal oedema follows **ascites; splenomegaly; prominent veins** are mainly present in the epigastrium and umbilical area.
2. IVC obstruction - Ascites with bipedal oedema may be present **but ascites follows oedema**. Prominent veins are seen over the abdomen, flanks and back; absence of splenomegaly.

3. SVC obstruction - Tortuous and prominent veins are present in the chest, upper arm with **facial puffiness**, chemosis of conjunctiva, and engorged and non-pulsatile neck veins; of en the patient is in respiratory distress.

Table 33 : Differentiation between portal hypertension and JVC obstruction

Features	Portal hypertension	IVC obstruction
1. H/O haematemesis or melaena	May be present	Absent
2. Sites of venous prominence	Epigastrium and periumbilical	Abdomen, flanks, back; lower limbs
3. Direction of flow	Away from the umbilicus	From below upwards
4. Ascites	Precedes oedema; ascites is commonly seen	Follows oedema; ascites is uncommon
5. Splenomegaly	Commonly present	Usually absent
6. Venous hum	May be present	Absent
7. Aetiology	Commonly cirrhosis of liver	Commonly abdominal or pelvic tumour, hypercoagulable states

•Visible vein' versus 'engorged vein' :

Sometimes, veins are visible normally in thin built persons (often in fair-skinned individuals) and is usually present at the skin level, i.e., flushed with the skin. But the engorged vein is a bit raised from the skin surface, which may be confirmed by tangential application of light. Palpate the vein lightly by index finger and draw your inference. Visibility of a vein does not mean that it is pathological—engorgement and moreover, *tortuosity indicates its pathological nature*.

Conclusion :

For obvious reason, it is preferable to choose a vein below the umbilicus for demonstration of venous flow in abdominal wall veins. Engorged and tortuous veins always indicate some underlying pathology.

Case 84

KAYSER-FLEISCHER (K-F) RING

What is this ?

The K-F ring is formed by the deposition of golden-brown pigment of copper in the Descemet's membrane of cornea in a patient suffering from Wilson's disease (hepato-lenticular degeneration).

Classical description :

The golden-brown pigment characteristically accumulate in the outer rim of cornea and the ring is broader superiorly and inferiorly than it is laterally and medially. The superior pole is first affected.

How the ring is detected ?

Many a time the ring can be seen in naked-eye examination. Examine the patient in sunlight, near a window or put a lighted torch in the eyes but slit-lamp examination by an ophthalmologist is usually necessary for confirmation. K-F ring is the clinical hallmark of Wilson's disease though their absence does not rule out the disease.

* Samuel A. K. Wilson (1878-1937) was an English neurologist.

Does it hamper vision ?

Never. It disappears after successful treatment.

Inference drawn, if K-F ring is detected :

The presence of K-F ring indicates that hepatic copper has been released in the circulation and

started damaging the brain. So, a patient of Wilson's disease with neurologic or psychiatric manifestations must have K-F ring in eyes. The commonest D/D at the bedside is arcus senilis.

Any other eye finding in Wilson's disease ?

There may be presence of sunflower cataract (15-20%).

Other conditions associated with K-F ring :

'K-F-like' ring may be seen in :

1. Cryptogenic cirrhosis,
2. Prolonged cholestasis due to any cause (e.g., primary biliary cirrhosis), and
3. Chronic biliary cirrhosis.

Types of hepatic involvement in Wilson's disease ;

In children, the liver is chiefly involved (**hepatic form**) though the clinical manifestations of excess of copper are rarely seen before the age of 6 years. The hepatic manifestations are in the form

1. Acute hepatitis,
2. Fulminant hepatitis (with haemolytic anaemia due to release of copper),
3. Chronic persistent and chronic active hepatitis,
4. Cirrhosis of liver, and
5. Asymptomatic hepatosplenomegaly.

* So, always search for jaundice and signs of hepato-cellular failure. Hepato-cellular carcinoma is very rare in Wilson's disease.

Neurological manifestations of Wilson's disease :

Patients presenting themselves after the age of 20 years usually suffer from neurological abnormalities (neurologic form). Neither there is any sign of sensory loss nor there are evidences of pyramidal tract involvement. The neurologic features (mainly of basal ganglia involvement) are :

- | | |
|----------------------------------|---------------------------|
| 1. Resting and intention tremor, | 6. Difficulty in writing, |
| 2. Chorea, athetosis, | 7 Grimacing, |
| 3. Rigidity, parkinsonism, | 8. Cognitive decline, |
| 4. Dysphagia, | 9. Drooling, and |
| 5. Slurred speech, silly smile, | 10. Dementia. |

* Three common movement disorders present are—dystonia, incoordination and tremor.

Psychiatric manifestations of Wilson's disease :

These are due to toxic effect of copper in the brain. Manifestations are in the form of :

- | | |
|--------------------------------|--------------------------|
| 1. Manic-depressive psychosis, | 3. Classic neurosis, and |
| 2. Schizophrenia, | 4. Bizzare behaviours. |

* Loss of emotional control (e.g., temper tantrums), hyperactivity and depression are common.

Investigations performed in Wilson's disease :

1. Low serum copper and ceruloplasmin (but can be normal).
2. 24-hours urinary copper is increased in the range of 100-1000 (µg).
3. Liver biopsy—affected patients have copper values > 200 µg/g of dry weight of liver ('gold standard for diagnosis).
4. Haemolysis as well as anaemia.
5. Genetic analysis (due to mutation in ATP7B gene localised to chromosome 13).

Normal values of plasma and urinary copper :

Serum copper : 70-140 mg/dl.

Serum ceruloplasmin : 18-35 mg/dl.

Urinary copper excretion : 20-50 Mg/day.

Plasma (free) copper is low but urinary copper is increased in Wilson's disease. It is an autosomal-recessive disease with low plasma ceruloplasmin, and high levels of copper in liver and brain.

* 10% patients may have normal ceruloplasmin level.

** Other rare manifestations of Wilson's disease are amenorrhoea, spontaneous abortions, osteoarthritis, haematuria, proteinuria and renal tubular acidosis.

*** Low serum ceruloplasmin level is also seen in fulminant hepatic failure, protein losing enteropathy and malabsorption. High level is found in pregnancy and biliary obstruction.

Management of Wilson's disease :

1. D-penicillamine (with pyridoxine)—Life-long therapy is needed with 1-1.5 g of penicillamine daily, orally in divided doses. Penicillamine is usually available as 250 mg capsule. Pyridoxine supplementation (25 mg/day) is beneficial. As it is a toxic drug and may worsen existing neurologic disease, it is not preferred as a first line drug.
 2. Elemental zinc as acetate (inhibits gastro-intestinal absorption of copper) — 50 mg, thrice daily. Zinc is the therapy of choice by many clinician.
 3. Trientine hydrochloride (1.2-1.8 g/day in 2 or 4 divided doses)—as a chelator.
 4. Low-copper diet, or avoidance of high-copper diet i.e., peanut, chocolate, mushrooms, liver, coffee, shellfish, legumes, nuts, dried beans should be avoided.
 5. Liver transplantation.
 6. Physiotherapy.
- * Potassium iodide (20 mg, four times daily), cobalt chloride and tetrathiomolybdate are other drugs used in Wilson's disease. Wilson's disease is potentially treatable.
- ** Siblings of the patient are screened for Wilson's disease and treated even if asymptomatic.

Side effects of penicillamine :

- | | |
|---|-------------------------------------|
| 1. Hypersensitivity reaction (rash, fever). | 6. SLE. |
| 2. Nausea, vomiting, mouth ulcers. | 7. Severe arthralgia. |
| 3. Agranulocytosis. | 8. Myasthenia gravis. |
| 4. Thrombocytopenia. | 9- Pemphigus. |
| 5. Nephrotic syndrome. | 10. Goodpasture's syndrome, rarely. |

It is mandatory to check routine blood count and urinary protein regularly when on penicillamine.

Use of penicillamine in clinical practice :

- | | |
|---|-------------------------------|
| 1. Wilson's disease. | 4. Scleroderma. |
| 2. Rheumatoid arthritis. | 5. Primary biliary cirrhosis. |
| 3. Chelating agent for copper, mercury, lead poisoning. | 6. Cystinuria. |

D/D of K-F ring :

(A) To an inexperienced eye, K-F ring may be confused with '**arcus senilis**'. Arcus is present in the periphery of cornea and is white in colour (usually starts in the lower pole of cornea). It is generally present above the age of 50 years, and represents deposition of phospholipid and cholesterol in corneal tissue (no significance in elderly people). If arcus is present before the age of 50 years, there is a possibility of underlying hyperlipidaemia (**arcus juvenilis**). It is not unduly associated with atherosclerosis, hypertension, myocardial infarction, diabetes mellitus or stroke. It does not affect vision.

(B) Rarely, corneal calcification (band keratopathy) which occurs at the lateral and medial margins of cornea in long-standing hypercalcaemia, e.g., in hyperparathyroidism, may be confused with K-F ring.

Conclusion :

Ability to incorporate radioactive copper (^{64}Cu) into ceruloplasmin differentiates Wilson's disease (little or no incorporation) from other copper-storage disorders (e.g., primary biliary cirrhosis).

It is a dictum that young patients with chronic liver disease showing mental abnormality, chorea, early ascites or haemolysis should always be screened for Wilson's disease, and the clinician should enquire for positive family history and search for K-F ring in that setting.

Case 85

PES CAVUS

Definition :

This is a fixed deformity of foot where both feet are more or less symmetrically high-arched i.e., there is gross exaggeration of the medial longitudinal arch of the feet. It may result from weakness of intrinsic muscles (lumbricals and interossei) which in turn may be due to poliomyelitis, spina bifida etc. Pes cavus may be associated with clawing of the toes. It is just opposite to pes planus (flat foot) deformity.

Method of demonstration at the bedside :

1. Ask the patient to stand on the floor and observe that only toes, webs and heel touch the floor surface, or
2. Take a foot-print on a white paper after painting the foot by lac-dye or any colouring agent, or
3. After immersing the feet in water, ask the patient to walk barefooted and watch the foot-prints on the floor.

Diseases associated with pes cavus (claw foot) :

Claw feet results from wasting of small muscles of feet due to any cause. The causes are :

- | | |
|-------------------------------|--|
| 1. Friedreich's ataxia. | 5. Spina bifida. |
| 2. Peroneal muscular atrophy. | 6. Cerebral palsy. |
| 3. Syringomyelia. | 7. Familial neuropathies e.g.. Refsum's disease. |
| 4. Poliomyelitis. | 8. Idiopathic. |

What is Charcot-Marie-Tooth disease ?

It is synonymous with peroneal muscular atrophy (a variety of hereditary sensori-motor neuropathy).

Situations where "Charcot's" name has been coined ?

1. Charcot-Marie-Tooth disease (hereditary polyneuropathy).
2. Charcot's biliary triad — Recurrent pain abdomen or biliary colic, fluctuating jaundice and intermittent fever with rigor in patients suffering from stone in CBD.
3. Charcot's intermittent hepatic fever— Fever due to cholangitis principally in the affection of intrahepatic biliary radicles.
4. Charcot joint or neuropathic joint (see page 484).
5. Charcot-Leyden crystals — Found in sputum of patients suffering from bronchial asthma or stool of patients suffering from acute amoebic dysentery.
6. Charcot's artery of cerebral haemorrhage (lenticulostriate branch of middle cerebral artery) — Usually associated with hypertensive cerebral haemorrhage.
7. Charcot's (cerebral) triad — Intention tremor, scanning speech and nystagmus seen in multiple sclerosis.
8. Charcot's vertigo — It is synonymous with 'cough syncope'.
9. Charcot zones — Hysterogenic zones in the human body.
10. Charcot-Neumann crystals — It is the phosphate crystals present in semen.

* Jean-Martin Charcot (1825-1893) was a physician in Hospital Salpetriere, Paris, France.

What is Friedreich's ataxia ?

It is an autosomal recessive form of spinocerebellar degeneration. The patient usually presents between the ages of 5 to 15 years, with clumsiness in walking and trunk ataxia. Gradually there is loss of proprioception and vibration sense with development of muscular atrophy, and loss of tone in the lower limbs. **Tendon reflexes are lost but plantar response remains extensor.** The degenerative process ascends upwards, and affects speech (dysarthria) and movements of the eyes (nystagmus). Optic atrophy (30%) may be seen. Kyphoscoliosis and pes cavus may be evident. It may be associated with diabetes mellitus and cardiomyopathy. The patient is wheel-chairbound by the age of 20. Diagnosis is made by positive family history and clinical findings.

- (i) Calculation—Serial 7-subtraction test (i.e., 100, 93, 86, 79, 72 ...) i.e., the arithmetic ability.
- (ii) Judgement — Ask the patient 'what will you do if you see a house set on fire, or a stamped and addressed envelope lying in the road in front of your house' ?
- (iii) Insight — Observe his awareness about the illness for which he has been admitted.
- (iv) Reasoning — Can he tell the difference between 'poverty and dishonesty', 'child and dwarf etc ?
- (v) Abstract thinking — Ask him the meaning of proverbs like 'all that glitters is not gold'.
- (vi) Attention — It is tested by. tapping the finger with repetition of a particular number.
- (vii) Concentration—Ask the patient to count backwards from 20 to 1.

$$* \text{ Intelligence quotient (IQ)} = \frac{\text{Mental age}}{\text{Chronological age}} \times 100.$$

** Mental retardation (assessed by IQ) : Mild (50-70), moderate (35-49), severe (20-34), and profound (< 20). Normal IQ is 90-109, whereas > 140 is regarded as near genius.

Mini-mental state examination (MMSE) :

This is a 'simple bedside test' to assess derangements in the higher functions (i.e., mental or intellectual functions), and is performed by Mental Status Questionnaire (MSQ). The results are scored numerically (the test takes 5-10 minutes and is scored out of 30). The functions tested are 1. Orientation 2. Registration 3. Attention and calculation 4. Recall, and 5. Language.

How do you classify the level of consciousness clinically ?

- (A) **Conscious** - Relates to the person who is alert, attentive and co-operative. Actually, it is a state of awareness of one's self and environment.
- (B) **Confused** - It denotes incapacity of the patient to think with customary speed and clarity. The patient is conscious but often talks irrelevantly.
- (C) **Drowsy** - The patient is sleepy but can be aroused easily by external stimulus.
- (D) **Stupor** - The patient who appears to be asleep can be aroused only by painful stimulus (pinching or supraorbital pressure).
- (E) **Semicoma** - When the patient responds only to internal stimulus (e.g., the patient may shake his body with fullness of urinary bladder) but not to external painful stimulus.
- (F) **Coma** - No response by internal or external stimuli, and the patient is deeply unconscious.

* The degree of coma or the level of consciousness is assessed now-a-days (as the terminology like drowsy, stuporose, semicoma are ill-defined) by **Glasgow coma scale (GCS)** but GCS can never be a substitute for thorough neurological examination. Circle the best scores from eye opening, motor response and verbal response, and compute the total.

Table 34 : Glasgow Coma Scale (clinical grading of coma)

EYE OPENING (E)	
Spontaneous	4
To speech	3
To pain	2
None	1
BEST MOTOR RESPONSE (M)	
Obeys commands	6
Localises pain	5
Flexion withdrawal	4
Abnormal flexion (decorticate posture)	3
Abnormal extension (decerebrate posture)	2
None (flaccid)	1
VERBAL RESPONSE (V)	
Oriented and converses	5
Confused and disoriented	4
Inappropriate words	3
Incomprehensible sounds	2
No response	1

N.B. : Coma scores = E+M+V : fully conscious =15, and deeply comatose = 3; scores >11 having 5-10% likelihood of death, while scoring 3 or 4 have 85% chance of dying

Diagnosis of spina bifida occulta :

Over the lower back, there may be :

1. Tuft of hair,
2. Dimpling of the skin,
3. Dilated vessels,
4. Fibrofatty tumour, or
5. Cicatricial thickening of skin.

Skeletal abnormalities in neurology :

1. Kyphoscoliosis,
2. Gibbus,
3. Pes cavus,
4. High arched palate (rare),
5. Various skull deformities, and
6. Short neck (cranio-vertebral anomaly).

Case 86**TESTS OF MEMORY****Different types of memory :**

1. Short term memory, immediate recall or rote memory.
2. Recent memory, and
3. Past memory, long term or remote memory.

What are the different components of memory ?

It may be arbitrarily divided into 4 parts :

1. Registration (defect found in manic states, senile dementia),
2. Retention (defect found in frontal lobe lesion, GPI),
3. Recall (defect found in epilepsy, Korsakoffs psychosis, post-traumatic states), and
4. Reproduction (defect found in CVA).

How to test the memory function at the bedside ?

Clinically 3 forms of memory are tested at the bedside :

- (A) **Short term memory** - memory which lasts upto 30 seconds, and is tested by,
 - (i) Whether the patient can repeat 'seven digits forwards' or 'five digits backwards' (digit span test).
 - (ii) Ask the patient to spell 'India' backwards.
 - (iii) Give him a telephone number and ask the number after 30 seconds or so.
- (B) **Recent memory** - It is the memory of events occurred within minutes, weeks, months and is an index of the severity of organic brain disease, which is usually tested by,
 - (i) Ask the morning news what he has read in the newspaper or seen on television.
 - (ii) Give him the names of three different objects and ask it after 3-5 minutes.
 - (iii) Ask him about the name of the visitors who met him last evening.
- (C) **Past memory** - This part of memory (events occurred years back) is relatively resistant to the effects of neurological insults. It is examined by,
 - (i) Ask the patient some important events in life e.g., date of birth, date of marriage etc.
 - (ii) Ask him the date of 'Independence day of India'.
 - (iii) Ask him the name of the first Prime Minister of India.

* In Wernicke-Korsakoff syndrome, the recent memory is lost with intact past memory whereas the past memory is affected in dementia (e.g., in the late stage of Alzheimer's disease).

What is dementia ?

It means loss of reasoning or more particularly, a deterioration of all intellectual and cognitive functions, without clouding of consciousness or disturbance of perception, and is due to chronic progressive degenerative disease of brain. In dementia, the patient is awake and alert but not aware.

* Two most important types of organic brain syndrome are delirium (acute) and dementia (chronic).

Causes of emotional lability or 'emotional incontinence' :

1. Cerebral atherosclerosis.
2. Multi-infarct dementia.
3. Pseudobulbar palsy.
4. Disseminated sclerosis.

Tests of 'intelligence' :

Intelligence is a total assessment of judgement, reasoning, arithmetic ability etc. and is tested by :

'Released reflexes' in dementia :

In infancy, developmental assessment is very often done by the presence (so called, 'primitive reflexes') or absence of these reflexes. In late infancy, these reflexes disappear as cortical control develops. In few pathological conditions in adults, these reflexes are released from the control of higher centres and thus, may be elicited. The examples are,

- (i) Dementia,
- (ii) Organic confusional states, and
- (iii) Sometimes in contralateral frontal lobe lesion.

The reflexes are :

1. **Grasp reflex** - Stroke the palmar surface of the patient's hand on its 'radial' aspect (region between the thumb and the index finger) by a distally moving stimulus. The patient immediately grasps the object and is unable to relax his grasp voluntarily. It is classically seen in contralateral frontal lobe lesion.
2. **Forced groping reflex**—sometimes, if the palmar surface ('radial' aspect) of patient's hand is lightly touched (patients' eyes are closed), the fingers close upon the object, and the hand and arm move towards the stimulus.
3. **Avoiding reflex** - It is the tendency of the patient's hand to move away from palmar stroking when an object is touched on the 'ulnar' side of the hand. It is commonly seen in contralateral parietal lobe disease.
4. **Palmo-mental reflex** - Scratch the thenar eminence with a pin, key or finger, and there will be puckering of the chin due to contraction of the ipsilateral mentalis muscle. It suggests contralateral frontal lobe lesion, pyramidal lesion or diffuse cerebral disease (may be seen normally in some elderly individuals). This reflex usually has no localising value.
5. **Sucking reflex** - The contact of an object with the lips or corner of the mouth evokes the movements of the lips, tongue and jaw concerned in sucking.
6. **Rooting reflex** - The lips follow the stimulating object when it is touched on the lips.
7. **Snout reflex** - A gentle tap of the examiner's knuckle against the centre of patient's closed upper lips will produce pouting of the lips due to reflex puckering of orbicularis oris.
8. **Glabella tap reflex (Myerson's sign)**- Normally after repeated tapping of the glabella by standing behind the patient, there are only three or four blinks before the response is inhibited. But in parkinsonism and senile dementia, the response goes uninhibited i.e.. a blink follows after each tap.

Common causes of loss of memory (dementia) :

- | | |
|--|-------------------------------------|
| 1. Transient — Following concussion, epilepsy. | 8. Hypothyroidism. |
| 2. Cerebral arteriosclerosis (multi-infarct dementia). | 9. Wilson's disease. |
| 3. Alzheimer's disease, Pick's disease. | 10. Wernicke-Korsakoff syndrome. |
| 4. Tumour affecting frontal lobe or corpus callosum. | 11. 'Normal pressure' hydrocephalus |
| 5. Alcoholism. | 12. Huntington's chorea. |
| 6. Chronic subdural haematoma. | 13. AIDS. |
| 7. Pellagra, vitamin B ₁₂ deficiency. | 14. Renal failure, hepatic failure. |

Case 87**SPEECH DISORDERS****What is speech ?**

It is the symbolic expression of thought process in spoken words or written words.

Types of speech disorders :

1. Aphasia or dysphasia (defect in higher centre with difficulty in language function).
2. Dysarthria (defect in articulation, commonly due to neuromuscular or muscular disorders resulting in impaired coordination of facio-lingual muscles)—slurring, mumbling etc.
3. Dysphonia (disorder of phonation due to abnormality in the vocal cord)—hoarseness, voice loss etc.

* Mutism means inability to speak while aphonia is inability to produce sounds.

Prerequisites before testing speech :

- | | |
|--------------------------------|--|
| 1. Level of consciousness, | 4. Intelligence level (education etc), |
| 2. Handedness (right or left), | 5. Deaf or not, and |
| 3. Mother language, | 6. Vision. |

* So, speech disturbance may be due to : 1. Deafness 2. Dementia/depression (severe) 3. Dysphasia (motor or sensory) 4. Dysarthria 5. Dysphonia.

Where the 'speech area' lies in the brain ?

In the right-handed person (90-95% of general population), it is present in the left hemisphere. In a left-handed person, it is present in the left hemisphere in 70% cases. In ambidextrous persons too, the left hemisphere is the dominant one. Dominant hemisphere also controls language and mathematical functions. The speech areas are distributed as follows :

- A) Motor or Broca's area (area 44) is situated in the posterior part of inferior frontal gyrus (spoken word) and in the posterior part of middle frontal gyrus (written word).
- B) Sensory or Wernicke's area (area 39 and 40) is situated in the posterior end of superior and middle temporal gyri.

Types of dysphasia or aphasia :**1. Expressive aphasia or nonfluent aphasia -**

- (i) Motor or Broca's aphasia — Commonest variety. The patient is unable to speak although there is no paralysis of tongue or lips.
- (ii) Agraphia — Inability to write although there is no paralysis of hand muscles.

2. Sensory or Wernicke's aphasia or fluent aphasia -

- (i) Word deafness — Though the patient can hear the sound, he is unable to analyse its meaning.
- (ii) Word blindness — The patient can see that something is written but he cannot recognise the words. His mother language appears as a foreign language to him.

3. **Global aphasia** - This is the combination of 1 and 2 i.e., there is defective comprehension as well as production of speech.

Speech evaluation at the bedside :

1. Spontaneous speech i.e., listen the articulation, content, comprehension and fluency.
2. Ask for naming simple objects like pen, coin, book etc.
3. Ask the patient to repeat words or phrases.
4. Instruct the patient to read aloud and to write a simple sentence.

How to examine an aphasic patient ?

1. Ask the patient his name. If he keeps mum, then it is the task of the physician to determine the variety of aphasia : motor or sensory type.
2. Now, write 'show me your tongue' on a white paper and show the paper to the patient. If he protrudes his tongue, then it is a case of expressive aphasia (comprehension is perfect and word blindness is not present) and if he does not protrude the tongue, probably you are dealing with a case of sensory aphasia or global aphasia. This test may be done by showing the examiner's protruding tongue to the aphasic patient.

What is the commonest cause of aphasia ?

Ischaemic cerebrovascular disease (CVA) is the commonest cause of aphasia. Cerebral haemorrhage causes aphasia less often than thrombosis because haemorrhage occurs deep in the white matter of the hemisphere, more often than in the cortex or subcortical regions. The patient with motor aphasia is often hemiplegic (right-sided hemiplegia) as most of the persons are right-handed.

Intracranial tumour is the commonest cause of aphasia during the first half of adult life.

What is nominal aphasia ?

The patient fails to name the common objects like pen, pencil, coin etc (produced in front of him) though he can tell the use of it. If he is told the actual name of the object, he immediately accepts it gladly.

Site of lesion — In between the angular gyrus and the posterior part of superior temporal gyrus.

What are the different dysarthrias ?

The normal requisites for production of syllables are—

- Articulation—labial, lingual and palatal muscles.
- Phonation—larynx.
- Resonance—nasopharynx.
- Along with normal respiration.

The different types of dysarthria are :

1. Spastic dysarthria — Difficulty in uttering the words but the words pronounced will be distinct. There is difficulty in pronouncing 'b', 'p' or 't'. It is commonly seen in pseudobulbar palsy.
2. Flaccid dysarthria — There is much difficulty in uttering the words and the uttered words will be slurred and indistinct. Often there is added nasal intonation ('egg' in pronounced as 'eng'). This type is commonly seen in bulbar palsy ('Donald Duck' quality), alcoholism etc.
3. Scanning or staccato speech — Read the section on 'Cerebellar disorder'.
4. Slow, monotonous speech without any fluctuation (i.e., lacking accents) — Seen in parkinsonism.
5. Lalling speech (voice) — Commonly seen in children, cretinism, Down's syndrome and mental retardation.
6. Stammering speech — Usually congenital in nature.

* In a patient of hemiplegia, dysphasia is due to involvement of dominant hemisphere, and dysarthria is usually due to UMN type of affection of VIIth and/or XIIth nerves.

** Ill-fitted dentures are very common cause of dysarthria.

*** VIIth nerve palsy = labial dysarthria; XIIth nerve palsy = lingual dysarthria.

What is dysphonia ?

Dysphonia is commonly due to laryngitis, tumour of the vocal cord or bilateral adductor paralysis (the voice becomes hoarse). In this disorder of vocalisation, the fault lies in the vocal cord.

How to diagnose hysterical aphonia ?

The hysteric aphonic patient (commonly a female) whispers but cannot talk aloud. Ask the patient to cough or give a painful stimulus. These two manoeuvres will definitely produce a sound.

* Echolalia—repetition of examiner's word, palilalia—repetition of terminal words of own speech, jargon aphasia—neologisms (new words) making no sense at all, alexia—inability to read, and perseveration—repeated use of particular words or phrases.

** Flaccid dysarthria, scanning and monotonous speech may be given in the examination.

Case 88

TREMOR

What is tremor ?

These are involuntary, repetitive, oscillatory movements of a part of the body around a fixed point due to alternate contraction and relaxation of groups of muscles with their antagonists.

How do you classify tremor clinically ?

(A) According to relationship with posture :

- | | |
|--|--------------------------|
| 1. Static tremor, | 3. Intention tremor, and |
| 2. Kinetic, postural or action tremor, | 4. Flapping tremor. |

(B) According to amplitude ;

1. Fine, and
2. Coarse tremor.

How will you examine a patient for tremor ?

At First, look for the presence of any **static tremor** (tremor at rest). This is classically seen in parkinsonism. In the hand, it is described as 'pill-rolling' or 'cigarette making' tremor. This very obvious tremor is characterised by,

1. The wrist and elbows are semiflexed, fingers flexed at MCP joints and extended at interphalangeal joints with abduction of the thumb. The oscillations of the fingers and thumb in two planes produce the 'pill-rolling' effect (flexion-extension at wrist and fingers, pronation-supination in forearm along with abduction-adduction of thumb and complex combinations).
2. It is a static tremor and does not increase at the goal point of action.
3. Coarse in nature (4-6/second).
4. More prominent in hands and can be controlled temporarily by willed movements.
5. Tremor is worse during anxiety or emotion, and is supposed to decrease on activity. It disappears during sleep.
6. It may involve the head, jaw, tongue, legs and even the trunk.

* Other features of parkinsonism like hypokinesia, rigidity, flexed posture and festinant gait will be present. There may be 'drum-beating' or 'bread cutting' tremor present in parkinsonism.

Now, look for the presence of any **kinetic** or **action tremor**. The patient is asked to outstretch his hands with separated fingers (a paper or thin piece of cardboard may be placed over the outstretched fingers) for the appearance of tremor, if present. This type of tremor is seen in anxiety, thyrotoxicosis, chronic alcoholism, by application of drugs (salbutamol, sympathomimetics, tricyclic antidepressants) or in familial tremor. Action tremor is characterised by,

1. Fine in nature (7-10/second).
2. Disappears at rest, and appears in precise and accurate movements.
3. May also be seen in tongue, lips and head.

* Familial tremor is often diminished by intake of alcohol.

Next the patient is examined for the presence of any **intention tremor**. This is classically seen in cerebellar disease, and rarely in severe parkinsonism. It is characterised by,

1. Coarse in nature (4-6/second).
2. Tremor appears at the goal point of an action (i.e., absent at rest and at the beginning of any movement). The patient is asked to hold a glass of water which is kept on the table or to do the 'finger nose test'. The movement becomes clumsy before he reaches the target.
3. Sometimes, there is tremor seen in head (titubation).

* Other signs of cerebellar disease like hypotonia, nystagmus, 'past pointing' etc. will be present.

Lastly, examine the patient for the presence of any **flapping tremor** (asterixis or bat's-wing tremor). This is elicited by three different manoeuvres :

1. By asking the patient to outstretch his arms with the hands extended at wrists and MCP joints, and to keep the limbs in this position for 20 seconds, when jerky forward movements of hand occurs every 2-3 seconds, or
2. By keeping the upper limb in bed with forearm fixed, the wrist is passively extended by holding the fingers. After few seconds, the pressure is released and the patient is instructed to keep the hand in the extended position (**classical method**). The response is described below..
3. Ask the patient to squeeze the physician's extended fingers—Flapping tremor is said to be present as indicated by repeated intensification and relaxation of the intensity of the squeeze ("milkmaid's grip").

Reponse— There is rapid flexion-extension movements at the MCP joints which are often accompanied by lateral movements of the digits and the hand ultimately falls (looks like 'good-bye'). This type of tremor is due to impaired inflow of joint and other afferent informations to the brainstem reticular formation which results in lapses in sustained posture, i.e., there is development of neuromuscular incoordination between flexor and extensor muscles. Flapping tremor is classically seen in,

- | | |
|--|------------------------------|
| (i) Hepatic precoma ('liver flap'), | (iv) Severe cardiac failure, |
| (ii) Renal failure, | (v) Hypnotic poisoning, and |
| (iii) Respiratory failure (carbon dioxide narcosis), | (vi) Toxic encephalopathies. |

* Flapping tremor is absent in hepatic coma.

What are the other sites you will examine next ?

1. THE TONGUE—The patient is asked to protrude the tongue out of the oral cavity for at least 1/2 minute (fasciculation in the tongue is seen when the tongue is relaxed in the floor of the mouth, and not protruded).
2. Head (and neck)—Titubation.
3. Jaw, lips, trunk or lower limbs.

* **One must examine tremor both in hands and tongue.**

Common causes of tremor :

(A) Fine (7-10/second) :

1. Anxiety, nervousness, fatigue.
2. Senile tremor.
3. Thyrotoxicosis.
4. Chronic alcoholism.
5. Drugs like salbutamol or terbutaline.
6. Familial (benign essential tremor).
7. General paralysis of insane (GPI).

(B) Coarse (4-6/second) :

1. Static tremor of parkinsonism.
2. Intention tremor of cerebellar disease or multiple sclerosis.
3. Flapping tremor of hepatic precoma, uraemia.
4. Sometimes, senile tremor.
5. Wilson's disease.

Types of tremor :

- | | |
|---|---|
| <ol style="list-style-type: none"> 1. Static. 2. Action. 3. Intention. 4. Flapping. | <ol style="list-style-type: none"> 5. Senile. 6. Physiological (anxiety). 7. Familial. 8. Hysterical. |
|---|---|

Commonest cause of tremor in clinical practice :

Physiological tremor due to anxiety, fear or nervousness.

Identification points (facies) for aetiological diagnosis of tremor :

See the face and observe for :

1. Exophthalmos—Thyrotoxicosis.
2. Flushed face with parotid swelling—Chronic alcoholism.
3. Masked facies—Parkinsonism.
4. Anxious face—Anxiety neurosis.
5. Very old person—Senile tremor or parkinsonism.
6. K-F ring with jaundice—Wilson's disease (asterixis).
7. Alae nasi moving (i.e., the patient is breathless) — Drug-induced (salbutamol).

Case 89

PROSIS

Definition :

Drooping of one or both upper eyelid.

What are the types of ptosis ?

1. Complete, and
2. Partial.

* Again, ptosis may be congenital or acquired.

** Recently, ptosis is divided into 5 types : blepharoptosis (eye surgery, old trauma), mechanical (oedema of upper eyelid), aponeurotic (cataract surgery), myogenic (myasthenia gravis, myopathies), and neurogenic (IIIrd cranial nerve palsy or Horner's syndroms).

How to recognise partial ptosis ?

By the narrowing of the palpebral fissure, when the clinician stands in front of the patient and the patient looks forward.

Common causes of ptosis :

1. Oculomotor nerve paralysis (usually complete, may be partial).
2. Horner's syndrome (always partial or pseudoptosis).
3. Tabes dorsalis (partial).
4. Myasthenia gravis (worsens as day progresses).
5. Myotonia dystrophica; ocular or oculo-pharyngeal myopathy.
6. Congenital (the upper eyelid is smooth and devoid of any cutaneous folds; and with compensatory head posture i.e., head tilts slightly backwards).

7. Snake bite (Elapidæ group).
8. Botulism.
9. Periodic paralysis.
10. Hysterical.
11. Oedema or tumour of upper eyelid, enucleation of eyeball.
12. Temporal arteritis.

* Congenital ptosis may be unilateral or bilateral. Acquired ptosis is usually **unilateral** in Illrd nerve palsy, Horner's syndrome, senility, traumatic and hysterical conversion reaction. It is **bilateral** in tabes dorsalis, myopathy or disorders of myoneural junction.

** Unilateral ptosis is a recognised finding in cluster headache, syringobulbia and cavernous sinus thrombosis.

*** 'Narrow palpebral fissure' results from ptosis and pseudoptosis, enophthalmos and blepharospasm.

What is the opposite phenomenon of ptosis ?

Retraction of the upper eyelid, i.e., spasm of the Muller's muscle in thyrotoxicosis due to sympathetic overactivity.

Nerve supply of the upper eyelid :

There are two muscles in the upper eyelid like,

1. Levator palpebrae superioris (LPS)—Supplied by Illrd cranial nerve.
2. Muller's (tarsal) muscle—Supplied by sympathetic trunk.

Test the power of LPS muscle or upper eyelid, or test for ptosis :

- (A) First step Stand in front of the patient face to face, and ask him to look upwards or elevate the upper eyelid voluntarily.
- (B) Second step—Now push down the frontal belly of occipitofrontalis muscle of forehead by your left hand (it is done to eliminate its elevating action on the upper eyelid). Again ask the patient to look upwards.
- (C) Third step—If the patient can elevate the upper eyelid, now you may apply little resistance by your right index finger over the upper eyelid and ask the patient to look upwards again (obviously with eliminating the action of forehead muscle). Compare with the other side.

* If the patient can not elevate the upper eyelid voluntarily, it is useless to do the next steps.

What is pseudoptosis ?

In a patient of Horner's syndrome, (i.e., damage to the cervical sympathetic fibres) though there is 'partial' drooping of the upper eyelid, the ptosis is often corrected on looking upwards voluntarily (due to intact Illrd nerve) and thus, it is known as pseudoptosis (i.e.. false ptosis).

Features of Illrd cranial nerve palsy :

1. Usually, it is unilateral 'complete ptosis' (as there is complete ptosis in one eye, patients do not complain of diplopia; patients are not able to open the affected eye voluntarily).
2. Lateral or divergent squint (due to unopposed action of lateral rectus). The patient is unable to move the eye upwards, downwards and medially.
3. Fixed (loss of light and accommodation reflex) and dilated pupil.

* Diplopia is complained by the patient as soon as the affected upper eyelid is elevated by fingers.

Components of Horner's syndrome :

Horner's syndrome results from damage to the cervical sympathetic fibres. It is the unilateral—

1. Pseudoptosis (diagnosed by narrowing of palpebral fissure; it is partial ptosis).
2. Miosis (constriction of the pupil, as cervical sympathetic supplies the dilator pupillae)—The pupil shows reaction to light.
3. Anhidrosis (involves ipsilateral half of the face and neck, front and back of upper chest, arm).
4. Enophthalmos (sympathetic supply maintains the tone of the eyeball)—shrunken eye.
5. Loss of ciliospinal reflex (in health, when the skin of the sides of neck is pinched, there is reflex dilatation of pupil and this reflex is abolished in Horner's syndrome).

* Johann F. Horner (1831-1886) was Professor of Ophthalmology, Zurich, Switzerland.

Trace the pathway of ocular sympathetic fibres :

1st order of neurones of sympathetic fibres descend from hypothalamus → brainstem → intermediolateral horn of spinal cord at C₈, T₁ level → 2nd order of neurones start and go upto superior sympathetic ganglion → 3rd order of neurones from there carried around carotid artery → ophthalmic division of Vth nerve nasociliary nerve → long ciliary nerve → oculopupillary fibres → supply dilator pupillae and Muller's muscle of upper eyelid.

Differentiate between Illrd nerve palsy and Horner's syndrome-induced ptosis ?**Illrd nerve palsy :**

1. Usually complete ptosis
2. Dilated pupil
3. Squint
4. Extraocular muscle palsy present

Horner's syndrome :

1. Partial ptosis
2. Constricted pupil
3. No squint
4. Extraocular muscles—normal

Can you test the Muller's muscle ?

No. It is an involuntary muscle.

Ptosis and pupil :

1. Ptosis and dilated pupil—Illrd nerve palsy.
2. Ptosis with constricted pupil—Horner's syndrome.
3. Ptosis with normal-sized pupil—myasthenia gravis, myotonia dystrophica, botulism (in majority), snake bite, and rarely in infarction or ischaemia of Illrd nerve (e.g., vasculitis, migraine or diabetes).

What is 'Tabetic fades' ?

This is the classical facies of tabes dorsalis and is characterised by :

1. Bilateral partial ptosis (as a result of paralysis of Muller's muscle),
2. Presence of compensatory wrinkling in the forehead (to overcome the partial ptosis),
3. Elevated eyebrows, and
- [4. Associated Argyll Robertson pupil],

* Increased furrowing in the forehead may be seen in any long-standing ptosis.

Speciality of ptosis in myasthenia gravis :

Ptosis may be unilateral or commonly bilateral, and appears towards the end of the day after repeated use of the upper eyelid (fluctuating ptosis with diurnal variation). Ptosis is corrected dramatically by the I.V injection of edrophonium hydrochloride (Tensilon test). There is **no alteration in the pupillary reflex or size of the pupil**. Sustained upward gaze for 2 minutes leads to increased ptosis.

Bedside tests for myasthenia gravis :

1. Maintain sustained upward gaze of eyes—observe the appearance of ptosis.
2. Counting sing—ask the patient to count upto 20 in one breath. The patient starts counting normally but the voice becomes slurred and unintelligible after counting few numbers due to development of muscle fatigue.
3. Ask the patient to close the eyes tightly and to do it repeatedly—eye closure becomes weaker after repetition.

* Myasthenia means fatiguable weakness which is worse on exercise, itself a neuromuscular junction disorder.

Dilatation (mydriasis) of pupil :

Constrictors of the pupil are supplied by parasympathetics (via oculomotor nerve), while the dilator fibres are controlled by sympathetic nervous system. Changes in the size of the pupil do not affect vision.

(A) Unilateral-

1. 3rd nerve palsy
2. Adie's pupil or myotonic pupil
3. Optic atrophy
4. Acute congestive glaucoma
5. Head injury

(B) Bilateral-

1. Childhood, anxiety, fear
2. Application of mydriatic (atropine)
3. Dhatura poisoning
4. Coma
5. Severe raised intracranial tension
6. Cerebral anoxia, death

Constriction (miosis) of pupil :

(A) Unilateral-

Horner's syndrome

(B) Bilateral-

1. Old age
2. Argyll Robertson pupil
3. Pontine haemorrhage
4. Organophosphorus or alcohol poisoning
5. Application of pilocarpine drops
6. Overdose of neostigmine
7. Iritis

Commonest cause of anisocoria (unequal pupils) is the application of mydriatic to one eye. Other causes are idiopathic or physiological (12% population), unilateral IIIrd nerve lesion, unilateral lesion in the sympathetic trunk, encephalitis, iritis, Holmes-Adie pupil etc.

** Point 3, 4, 5, morphine or barbiturate poisoning, heat stroke may produce **pin-point pupil**.

*** Irregular pupils are due to coloboma, neurosyphilis, following eye operation. Sometimes, it is seen in normal healthy subjects.

How to differentiate between patients with pin-point pupil (point 3 and 4 above) :

- (A) Pontine haemorrhage—There is coma, hyperpyrexia and long tract signs (i.e., signs of pyramidal tract lesion).
- (B) Organophosphorus poisoning—It is diagnosed by history, absence of long tract signs and signs of respiratory depression. The patient may be unconscious but there is absence of pyrexia.

Causes of Horner's syndrome :

The site of lesion may be in,

1. Cerebral hemisphere—Massive infarction of one hemisphere.
2. Brain stem—Lateral medullary syndrome (vascular), multiple sclerosis (demyelinating), pontine glioma (SOL), syringobulbia, encephalitis.
3. Spinal cord (lower cervical)—Syringomyelia, glioma, ependymoma.
4. Spinal cord (T root lesions)—Cervical rib. trauma, Klumpke's paralysis, aortic aneurysm.
5. Cervical sympathetic chain—Pancoast's tumour, enlarged lymph nodes, mediastinal mass, thoracic surgery, trauma.
6. Carotid artery—Thrombosis, spasm in migraine (temporary Horner's syndrome).
7. Others—Congenital, idiopathic.

* Congenital Horner's syndrome have ipsilateral facial hemiatrophy and light colouration of iris.

What is the size of normal pupil ?

It varies in between 3-5 mm. If < 3mm, it is called miosis and if > 5 mm, it is known as mydriasis. Pupillary size 1 mm or less is known as pin-point pupil. Normally pupils are 'round and regular' in outline, centered in the iris, and equal in size.

Argyll Robertson pupil :

(A) Characteristics :

- a) Small, irregular and unequal pupil; often depigmented.
- b) Usually bilateral involvement but more marked on one side.
- c) Accommodation reflex present but light reflex lost.
- d) Dilates slowly to mydriatics.

(B) Site of lesion : Peri-aqueductal grey matter (pretectal region of mesencephalon). Probably there is damage of both colliculo-nuclear and sympathetic fibres.

(C) Clinical associations :

- | | |
|--------------------------------------|----------------------------|
| 1. Neurosyphilis (classically seen). | 5. Brainstem encephalitis. |
| 2. Diabetes mellitus. | 6. Amyloidosis. |
| 3. Wernicke's encephalopathy. | 7. Pinealomas. |
| 4. Multiple sclerosis. | |

What is 'reversed' Argyll Robertson pupil ?

The pupil reacts to light but not to accommodation reflex. This rare phenomenon may be seen in post-encephalitic parkinsonism and diphtheria.

Myotonic or Adie's pupil :

It is characterised by delayed or absent pupillary reaction to light or to accommodation, but once constricted the pupil dilates very very slowly. The lesion is in ciliary ganglion.

* **'Hippus'** is rhythmic constriction and dilatation of pupil, either spontaneously or in response to light. It is not of much clinical importance.

Describe the pathway of light reflex :

Light falls on retina-----optic nerve-----optic chiasma----- optic tract----- superior colliculi at mid-brain (pretectal area)-----2nd order of neurone starts from here and goes to the Edinger-Westphal (E.W) nucleus bilaterally----- preganglionic fibres go to the ciliary ganglion through the oculomotor nerve and its branch to the inferior oblique muscle----- postganglionic fibres from ciliary ganglion supply the sphincter pupillae and ciliaris muscle through short ciliary nerve.

Describe the pathway of accommodation reflex :

Light falls on retina-----optic nerve----- optic chiasma----- optic tract-----lateral geniculate body----- optic radiation-----calcarine cortex of the occipital lobe (visual cortex)----- frontal eye field area or 'area 8' through superior longitudinal association tract----- from here cortico-mesencephalic fibres descend to the oculomotor nuclei at midbrain as follows :

1. Nucleus for medial rectus—produces medial convergence of eyeball.
2. E.W nucleus—produces miosis.
3. Nucleus of Edinger—contraction of ciliaris muscle and thus, anterior convexity of lens increases.

* From oculomotor (E.W) nucleus, the pathway for accommodation reflex is the same as light reflex.

How to test for light reflex ?

When light falls on one eye, there is constriction of both the pupils. The pupillary reaction on the stimulated side is called 'direct' light reflex, and the pupillary constriction of the other eye is known as 'consensual' light reflex. The method of examination goes like below :

1. Pencil torch is used with good power of illumination.
2. The patient is asked to look straight forward at a distant object and the light is thrown suddenly in one eye **from sides** (to avoid the accommodation reflex). The pupil constricts briskly.
3. For direct light reflex, opposite eye should be closed by the hollow of the other palm. Direct light reflex should be tested preferably in a dark room.
4. For consensual light reflex, one hand of the patient is placed over the nose (like a curtain) to prevent the spillage of light to the other eye. Both the eyes are kept open. When light falls on one eye, observe the pupillary constriction of the other eye (i.e., consensual light reflex).
5. The afferent pathway—optic nerve, efferent pathway— oculomotor nerve, centre—ciliary ganglion, response—constriction of both the pupils.

* For both direct and consensual light reflexes, each eye is tested separately.

Mechanism of consensual light reflex :

As the fibres leaving one optic tract enter both the E.W nucleus, there is consensual light reflex.

How to test for accommodation reflex or convergence reflex ?

The patient is first allowed to look at a distant object and then to ask him to look at examiner's finger quickly which is placed near his nose (e.g., at 10 cm from the eyes). The responses are,

1. Miosis (bilateral).
2. Medial convergence of the eyeball (bilateral).
3. Increased anterior convexity of lens (not clinically evident).
4. Enophthalmos (not clinically evident).

Afferent pathway—optic nerve, efferent pathway—oculomotor nerve, centre—oculomotor nuclei at midbrain.

How will you test the integrity of intraocular muscles ?

By light (direct and consensual) and accommodation reflex.

Identification points :

In the presence of ptosis :

1. **Miosis indicates Horner's syndrome** and **mydriasis indicates oculomotor palsy**. Ptosis due to Horner's syndrome is partial.
2. Lateral squint with complete ptosis indicates oculomotor palsy.

3. Wrinkling of the forehead points towards long-standing ptosis.
4. Abnormal fatiguability of muscles after repeated use indicates myasthenia gravis.
5. Facies characterised by ptosis, 'hatchet face' with 'swan-neck', frontal baldness, cataract with atrophy of temporalis and sternomastoid muscles indicate myotonia dystrophica.

Case 90

SQUINT

What is squint ?

In squint, abnormality of ocular movement is such that the visual axes do not meet at the point of fixation or it is a failure of the normal coordination of the ocular axes. 'Manifest squint' is noticeable when both the eyes are kept open while 'latent squint' is detected only by covering one eye.

Types of squint :

Squint or strabismus is of two types :

1. Concomitant (non-paralytic)—There is no paralysis of extraocular muscles.
2. Paralytic—Due to weakness of one or more of the extraocular muscles.
 - (i) Divergent squint—Due to paralysis of MR (medial rectus).
 - (ii) Convergent squint—Due to paralysis of LR (lateral rectus).

* Upward or downward paralytic squint is rare.

Nerve supply of extraocular (extrinsic) muscles :

- a) Lateral rectus (LR)—VIth cranial nerve (abducens).
- b) Superior oblique (SO)—IVth cranial nerve (trochlear).
- c) Other four muscles (MR, IO, SR, IR)—IIIrd cranial nerve (oculomotor).

* IO—inferior oblique, SR—superior rectus, and IR—inferior rectus.

** **Intraocular (intrinsic) muscles**—(i) Sphincter pupillae and ciliaris—Supplied by IIIrd cranial nerve, (ii) Dilator pupillae— Supplied by sympathetic trunk.

Testing of IIIrd, IVth and VIth cranial nerves :

It is customary to test these three nerves at a time under the following headings :

- I. Upper eyelid :
 - a) Ptosis, and
 - b) Lid retraction (due to sympathetic overactivity).
- II. Position of the eyeball at rest :
 - a) Squint,
 - b) Exophthalmos,
 - c) Enophthalmos,
 - d) Hypertelorism (may be associated with intracranial congenital abnormalities), and
 - e) Conjugate deviation.
- III. Movement of the eyeball :
 - a) Voluntary, and
 - b) Involuntary—Nystagmus.
- IV. Pupil :
 - a) Size b) Shape, and c) Reaction :
 - (i) Light reflex,
 - (ii) Consensual light reflex,
 - (iii) Accommodation reflex.

N.B. : **To summarize.** test for 1. LPS muscle and ptosis, 2. Movement of each extraocular muscle,

3. Pupil with reflexes, and 4. Nystagmus.

* Lid retraction, enophthalmos, hypertelorism have no relation with IIIrd, IVth or VIth nerve palsy.

** One should search for arcus senilis (may have atheromatous cerebral vessels) and K-F ring (neurologic form of Wilson's disease) while examining the cornea and iris.

*** Retraction of upper eyelid (thyrotoxicosis, Claude Bernard syndrome, big eye globe, scar of upper eyelid, short tendon of LPS) and retraction of lower eyelid (ectropion, proptosis, early stage of Bell's palsy, ocular myopathy) should be carefully looked for.

How to test for voluntary movements of the eyeball ?

It is important to remember that the recti are elevators and depressors alone when the eye is in abduction, and obliques act similarly (elevation and depression) when the eye is in adduction.

Stand in front of the patient and ask him, not to move his head. He is then instructed to follow your finger which is kept at a distance of 60 cm, and placed sequentially from position 1 to 7 described below. Ask the patient to report for double vision (diplopia).

Position of examiner's finger

1. Above his head in the midline
2. Below his head (finger kept at the level of his chest) in the midline
3. Laterally to the left
4. Laterally to the right
5. Above his head but placed laterally
6. Below his head but placed laterally
7. Straight ahead

No. 5 and 6 are done on both the sides.

Muscles working

1. SR and IO (both eyes)
2. IR and SO (both eyes)
3. LR (left eye) and MR (right eye)
4. MR (left eye) and LR (right eye)
5. SR (same eye) and IO (opposite eye)
6. IR (same eye) and SO (opposite eye)
7. All extraocular muscles

According to few clinicians, it is better to test individual muscle of each eye by an H pattern of movement.

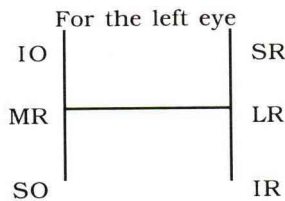


Fig. 3 : Testing for voluntary movements of eye

Function of each muscle :

- LR — abduction
- SR — elevator in abduction
- SO — depressor in adduction
- IO — elevator in adduction
- IR — adduction
- IR — depressor in abduction

Different voluntary movements of the eyeball :

The three components of voluntary movements are :

1. PURSUIT (following movements)—The movements of the eyeball described above i.e., slow and smooth eye movements following a target.
2. SACCADIC (searching movements)—These are rapid, programmed, conjugate fixation movements. Ask the patient to focus his eyes rapidly to the ceiling fan, then to the window and next to the switch-board and so on.
3. REFLEX OCULAR MOVEMENTS—To see whether both the eyes are moving simultaneously or not, on examiner's command i.e., the conjugate deviation.

* LMN lesion of IIIrd, IVth or VIth cranial nerve (i.e., nucleus or nerve lesion) produces paralysis of individual or group of eye muscles. UMN lesion leads to paralysis of conjugate gaze rather than individual muscle paralysis.

Table 35 : Clinical differentiation between two types of squint

Features	Paralytic	Non-paralytic
1. Onset	Acquired later in life	Present since childhood
2. Movements	Restricted	Good in all directions
3. Diplopia	Present	Absent
4. Vision	Normal in both eyes	Deviating eye has defective vision

Diplopia results from loss of parallel visual axes, and is commonly due to IIIrd, IVth, VIth nerve palsy, myasthenia gravis and myopathies (e.g., thyrotoxicosis).

Why there is no diplopia in non-paralytic squint ?

In case of paralytic squint, the image on the paralysed side does not fall on the macula and thus diplopia or double vision occurs; whereas in non-paralytic squint, the image formed by the defective eye is either rejected or suppressed by the occipital cortex and thus, there is no diplopia. Diplopia is maximum if the eye is moved in the direction of weak muscle.

Remember, uniocular diplopia is a rare phenomenon and mostly occurs due to eye disorders like incipient stage of cataract, subluxation of lens, peripheral iridectomy, astigmatism and keratoconus.

Causes of IIIrd, IVth and VIth cranial nerve palsy :

(A) Isolated oculomotor (IIIrd cranial nerve) palsy :

- | | |
|---|---|
| 1. Intracranial aneurysm arising near circle of Willis. | 5. Ophthalmoplegic migraine. |
| 2. Tuberculous meningitis. | 6. Neoplastic diseases (e.g., pituitary SOL). |
| 3. Diabetes mellitus. | 7. Multiple sclerosis. |
| 4. Traumatic. | 8. Vascular disease (e.g., vasculitis). |
| | 9. Congenital. |

(B) Isolated trochlear (IVth cranial nerve) palsy ;

Isolated lesion is rare e.g., in trauma (head injury), diabetes mellitus etc. It is almost always involved with IIIrd or VIth nerve paralysis in multiple sclerosis, Wernicke's encephalopathy.

(C) Isolated abducens (VIth cranial nerve) palsy :

1. Increased intracranial tension (it is a 'false localising sign'; VIth nerve is commonly affected due to its long intracranial course).
2. Diabetes mellitus.
3. Traumatic.
4. Brain tumour (SOL).
5. Basal meningitis or encephalitis.
6. Gradenigo's syndrome (unilateral headache, VIth nerve palsy with mastoid infection).
7. Lesion in cavernous sinus (aneurysms, meningioma).

* Combined IIIrd, IVth and VIth nerve palsies are common in upper brainstem stroke, cavernous sinus thrombosis, superior orbital fissure syndrome (i.e., Tolosa-Hunt syndrome) and diabetes mellitus.

Symptoms due to isolated IVth nerve palsy :

in superior oblique palsy, the patient feels difficulty in going downstairs or reading newspapers. The patient overcomes this difficulty by tilting his head (head tilt) slightly to opposite direction.

What is ophthalmoplegia ?

1. Internal ophthalmoplegia Occurs as a result of paralysis of sphincter pupillae and ciliaris muscles due to IIIrd nerve palsy. The pupil will be dilated and fixed, i.e., there will be absence of direct and consensual light reflex as well as accommodation reflex.
2. External ophthalmoplegia — Occurs due to IIIrd, IVth or VIth nerve palsy (e.g., cavernous sinus thrombosis or metastasis at orbital apex); and extraocular muscle paralysis due to myasthenia gravis, ocular myopathy and exophthalmos.
3. Internuclear ophthalmoplegia (INO) — Lesion in the medial longitudinal fasciculus (MLF) anywhere in between the midbrain and the pons will produce difficulty in adduction in one eye and nystagmus in the abducting eye, and the side of lesion is on the side of impaired adduction. It is commonly due to multiple sclerosis.

Points to note during optic nerve examination :

1. Acuity of vision (measures efficacy of the central or macular vision),
2. Field of vision (assess the function of central and peripheral retina),
3. Colour vision (assess the function of retinal cones and optic nerve), and
4. Fundoscopy (to examine optic nerve head and fundus).

According to some clinicians, pupillary reflexes are within optic nerve examination.

Neuroanatomy of optic nerve :

Retina -> optic nerve optic chiasma (here, nasal fibres from each retina, representing the temporal

field, decussate. Temporal fibres from each retina remain on the same side) → optic tract → lateral geniculate body (some preganglionic fibres project to superior colliculi at midbrain) → optic radiation projects through posterior limb of the internal capsule → to calcarine cortex of the occipital lobe. Fibres carrying the upper and lower visual fields pass respectively through the white matter of temporal and parietal lobe; the right half of the visual field is represented in the left calcarine cortex and vice versa.

How to test for visual acuity or acuity of vision ?

It is a test of 11th cranial nerve and it denotes the macular function. The clinical method is .

1. First examine the eye for any local pathology i.e., corneal opacity etc.
2. One eye is tested at a time (other eye is closed by the hollow of (lie palm).
3. Ask the patient to count the beams in the ceiling or blades in the fan (distant object testing).
4. Now ask for finger counting (near object testing).
5. If finger counting is not possible, put a lighted torch in the eye and examine for PL/PR (perception of light / projection of rays). Perception of light is absent in total blindness.

* Ideally, distant vision is tested by SNELLEN'S TEST TYPE CHART and near vision by JAEGER'S CHART. The patient should wear the spectacles (if any) during the bedside test.

Normal visual acuity by Snellen's chart is $\frac{6}{6}$; in clinical practice, loss of visual acuity is commonly due to cataract, refractive error, corneal and vitreous opacities.

How to test for colour vision ?

The primary colours are red, green and blue. Roughly, it can be assessed by asking the patient the colour of his shirt or pant, room-wall, ceiling fan, bed-cover etc. The patient with defective colour vision can not identify the colours correctly.

Ideally it should be assessed by ISHIHARA'S CHART (pseudo-isochromatic plates). The commonest defect in colour vision is red-green anomaly (X-linked recessive), acquired defect may be seen after use of ethambutol or chloroquine. Total colour blindness is rare.

How to test for field of vision ?

'Field of vision' is the field within which all objects are visible when the vision is fixed to a particular object present in that field. This is tested by '**confrontation perimetry**'.

1. First test the acuity of vision.
2. Seat opposite the patient (face to face) at the same level; distance between the patient and the examiner will be one meter. The examiner should have a normal visual field.
3. If the left eye is to be tested, ask the patient to cover his right eye with the hollow of his right palm; ask him to look straight at your right eye and not to take his own eye off yours (the patient should not move his head). Cover your left eye with your left palm and look steadily at the patient's left eye (in this way of testing, slight movement of patient's eye can be easily detected).
4. Now place your right hand (the test object is physician's finger or ideally a large red hat-pin) midway between patient's face and your own. It is better to *move the finger during testing* and *the finger is brought from the periphery to the centre*. Ask the patient whether he can see the movement of the finger,
5. In this way, the hand (finger) is moved in various directions—i.e., upper and lower, nasal and temporal quadrants of the visual field. Bring the test object from the periphery into the field of vision in a curved fashion. Use your own field of vision for the purpose of comparison. Lastly, test the field of the right eye of the patient.

* In a non-cooperative patient, a shiny object is moved from the periphery to the centre and ascertain whether the patient is able to see it, or move your hand quickly towards patient's face and observe the reflex blinking of both eyes (menace reflex), as confrontation method is not possible here (this method can also be applied in a patient who is unable to sit in the bed). Ideally, the field of vision is mapped by using a BJERRUM'S SCREEN PERIMETER (perimetry). One may detect the physiological blind spot by confrontation method. Preferably remove both the examiner's and patient's spectacles (if any) while testing the field by confrontation method.

** 'Tubular or tunnel vision' (concentric narrowing of the field of vision) is characteristic of terminal glaucoma, advanced retinitis pigmentosa, quinine amblyopia and hysteria,

*** 'Cortical blindness' is a feature of vasospasm of posterior cerebral artery, anoxic encephalopathy, hypertensive encephalopathy, subarachnoid haemorrhage and migraine.

Causes of papilloedema, optic atrophy and optic neuritis :

- **Papilloedema (oedema or swelling of the papilla, i.e., the optic disc) :**

1. Increased intracranial tension (CVA, SOL, cerebral oedema, meningitis, hydrocephalus).
2. Malignant hypertension.
3. Circulatory block (in CSF).
4. Benign intracranial hypertension (steroid withdrawal, hypervitaminosis A).
5. Cavernous sinus thrombosis.
6. SVC syndrome.
7. Systemic disorders (vasculitis, chronic hypercapnia, COPD, hyperviscosity syndrome).
8. Miscellaneous Guillain Barre syndrome, pseudotumour cerebri, severe anaemia, central retinal vein occlusion, disc infiltration (leukaemia, sarcoidosis).

* Papilloedema is usually bilateral. Unilateral papilloedema is seen in central retinal vein occlusion, Foster-Kennedy syndrome, lymphomatous/leukaemic deposits in one eye, and early stage of bilateral papilloedema.

- **Optic atrophy :**

- | | |
|---|---|
| 1. Multiple sclerosis (Devic's disease). | 5. Ischaemic optic atrophy. |
| 2. Toxins—Tobacco, methyl alcohol, ethambutol, chloroquine etc. | 6. Head injury. |
| 3. Syphilitic optic neuritis. | 7. Retinitis pigmentosa, chorioretinitis. |
| 4. Glaucoma. | 8. Leber's optic atrophy (familial). |
| | 9. Friedreich's ataxia. |

Optic atrophy is of two types. **Primary optic atrophy** is secondary to optic neuritis, and **secondary optic atrophy** develops from papilloedema/papillitis. A third variety of optic atrophy is 'consecutive' optic atrophy resulting from primary retinal diseases (e.g., retinitis pigmentosa, chorioretinitis and central retinal artery occlusion).

- **Optic neuritis :**

1. Multiple sclerosis, Devic's disease.
2. Vitamin B₁ or B₁₂ deficiency.
3. Meningitis, encephalitis, tuberculosis, syphilis.
4. Toxins : tobacco, alcohol (ethyl or methyl); drugs : INH, ethambutol, quinine.
5. Diabetes mellitus, vasculitis.

* On **fundoscopy** Optic disc (colour, shape, blood vessels, physiological cup, margin), retinal blood vessels, macula and periphery of retina are seen.

The visible part of retina is known as fundus'. Fundoscopy should be routinely done irrespective of patient's complaints.

Fundoscopy in different conditions :

- (I) Papilloedema—the disc is pink (hyperaemic), blurring of disc margin (nasal margin first), cessation of normal venous pulsation, obliteration of physiological cup, disc engorgement with dilated vessels, and small haemorrhages surround the disc.
- (II) Optic atrophy the disc looks pale (due to loss of nerve fibres); the margin is sharp in primary atrophy while it is blurred in secondary atrophy. The arteries are attenuated in primary variety. No change in retina in primary variety but few exudates and haemorrhages may be seen in retina in secondary variety.
- (III) Retrobulbar neuritis—though the patient is blind, the disc is absolutely normal.

Precautions adopted in testing olfactory nerve :

Ideal test objects (non-irritating substances) are—Oil of peppermint, oil of cloves, tincture of asafoetida or oil of lemon. But in the examination, the students should test it by common bedside familiar substances like soap, tooth paste, fruit, scent etc. Irritating substances like ammonia stimulate trigeminal nerve too, and are better to be avoided.

Precautions :

1. Exclude local changes first (i.e., nasal catarrh, patency of the nostrils).
2. Examine each nostril separately after occluding other nostril by digital pressure.
3. Avoid irritating substances like ammonia (they stimulate the trigeminal nerve and may interfere with interpretation of smell).
4. Eyes remain closed during testing. Ask the patient to identify the substance with two sniffs.

5. The test is usually carried out with two or more substances.

* *Anosmia* is usually due to lesion in the olfactory bulb; *parosmia* or perverted smell (pleasant odours seem offensive) results from hysteria; *olfactory hallucination* (usually of unpleasant nature) is found in temporal lobe epilepsy. *Cacosmia* (foul smell) is found in atrophic rhinitis. Commonest cause of bilateral anosmia is coryza. Other causes of anosmia are local disease of cribriform plate, basal meningitis, subarachnoid haemorrhage, zinc deficiency and ageing.

** **Neuroanatomy** : The olfactory and optic nerves actually made up of central nervous tissue rather than peripheral nerves. The olfactory nerve carries the sense of smell. The axons of the bipolar sensory cells in the olfactory epithelium pass through cribriform plate → to reach the second order of neurones in olfactory bulb → the olfactory bulb neurones project to the olfactory centres (uncus and parahippocampal gyrus).

Identification points (squint) :

1. First determine the type i.e., paralytic or non-paralytic squint by testing the voluntary movement of the eyeball (test all extraocular muscles).
2. Ask for diplopia and the duration of squint.
3. See the pupil. If there is dilatation of pupil (and ptosis) in the paralysed eye, the squint is due to oculomotor palsy (divergent squint due to unopposed action of lateral rectus).
4. If the patient complains of diplopia (obviously paralytic squint), usually the outer image is a false one and it disappears on covering the affected eye.

Case 91

NYSTAGMUS

What is nystagmus ?

These are rhythmic, involuntary and jerky movements (oscillations) of the eyeball when they are fixed on an object. Nystagmus signifies the disturbance in ocular posture.

How to test for nystagmus ?

1. Examine the patient under good illumination. At first, test the power of extraocular muscles to eliminate their weakness or paralysis (in that situation, nystagmoid movements will appear).
2. Now ask the patient to look forward. Observe for pendular nystagmus.
3. Ask the patient to fix his head or fix the patient's head by one of your hands.
4. Now the patient is asked to fix his vision towards your index finger (held at about 60 cm distance) which is kept at least for 5 seconds laterally to the right side and then to the left side in turn (your finger must be placed within the binocular vision of the patient; do not go for extreme abduction to avoid nystagmoid movement.). It is the examination of nystagmus in the horizontal plane.
5. Next place your finger above and below his head to examine nystagmus in the vertical plane.
6. Observe nystagmus in both eyes.

How will you describe the nystagmus (if present) ?

Nystagmus is usually described under these 6 points.

1. **Pendular** (equal horizontal movement on each side like the swing of a pendulum of wall-clock) or **jerky** (amplitude is more in any one side. i.e. it has a fast and a slow component).
2. **Direction** of the nystagmus—The direction of the faster component of the oscillations is the 'direction' of the nystagmus.
3. **Plane in which nystagmus is being tested** (i.e., direction of gaze which evoke nystagmus)
 - (i) Horizontal, and
 - (ii) Vertical.
4. **Plane in which oscillatory movements of nystagmus is occurring**
 - (i) Horizontal (to and fro movement of the eyeball in the horizontal plane),
 - (ii) Vertical (up and down movement of the eyeball in the vertical plane), and
 - (iii) Rotatory (oscillatory movement of the eyeball is rotatory in character).
5. **Amplitude—**
 - (i) Fine (< 1mm),

- (11) Medium (1-3 mm), and
 - (iii) Coarse (> 3 mm).
6. **Grading** (discussed below).

Causes of pendular nystagmus :

Pendular nystagmus is almost always binocular and horizontal, any sometimes associated with head-nodding. **Pendular nystagmus appears when the patient looks forward.** The common causes are,

- (i) Miners nystagmus (who works in dim light of coal-mines for long duration).
- (ii) Errors of refraction—the ocular fixation is defective.
- (iii) Amblyopia (if present in early life, impairs fixation of vision) e.g., high infantile myopia.
- (iv) Familial nystagmus, albinism, retinitis pigmentosa.

Causes of jerky nystagmus :

Jerky nystagmus is always found on fixation of the gaze laterally.

- (i) Cerebellar nystagmus Direction (fast phase) of the nystagmus is towards the side of lesion.
- (ii) Labyrinthine nystagmus—Direction (fast phase) of the nystagmus is towards the opposite side of lesion.
- (iii) Brainstem lesion—Very often 'rotatory' e.g., in brainstem glioma, multiple sclerosis etc.
- (iv) Upper spinal cord lesion—Vertical nystagmus e.g., Arnold-Chiari malformation.
- (v) Toxic nystagmus—Induced by drugs like benzodiazepines, phenytoin, phenothiazines, barbiturates and sometimes in alcoholics.

* vertical nystagmus : Downbeat (i.e., fast phase downwards and slow phase upwards) in cranio-vertebral junction abnormality e.g., Arnold-Chiari malformation, and upbeat nystagmus (i.e., fast phase upwards and slow phase downwards) in lesion of cerebellar vermis.

What is positional nystagmus (Dix-Hallpike's test) ?

Method—The patient is asked to lie in supine position with his shoulders at the end of the couch (with eyes open). The projecting head of the patient is supported by the examiner's hand. The head is now fully extended (30° from horizontal plane) and rotated (30°) to one side. Intense vertigo with nystagmus appears (affected side down) in vestibular dysfunction. It is the rough assessment of the vestibular function of VIIIth cranial nerve. After a short interval, the test is repeated with the head extended and rotated to the other side. Positional nystagmus is also seen in posterior fossa tumour.

What is optokinetic nystagmus ?

It is commonly seen when an individual looks out of the window of a moving train. There is a slow and a fast phase in it. It is a physiological nystagmus and its absence indicates parietal lobe lesion.

What is see-saw nystagmus ?

In this condition, one eye raises up and turns in, while the other eye falls and turns out. It is common in parasellar tumour.

What are nystagmoid movements ?

These are rhythmic movements of the eyeball when the eyeball is extended laterally beyond the limit of binocular vision. **Brief duration and irregularity** of these movements distinguish them from the true nystagmus. There are usually 4 or less oscillations in the nystagmoid movement. Often this movement is monocular.

What is the grading of nystagmus ?

For example :

Grade 1— Nystagmus with fast component to left, when the patient looks towards the left.

Grade 2— Nystagmus with fast component to left, when the patient looks straight ahead.

Grade 3— Nystagmus with fast component to left, when the patient looks towards the right.

This example stands for a left-sided cerebellar hemispheric lesion or a right-sided labyrinthine lesion.

What do you mean by nystagmus to the left ?

It does not refer to the direction of gaze in which nystagmus occurs but it indicates the direction of last component to the left (i.e., 'direction' of the nystagmus).

* Nystagmus is routinely tested during examination of the IIIrd, IVth and VIth cranial nerves. Always fix the head of the patient. Test for nystagmus in different planes.

Few other abnormal involuntary movements of eyes :

1. Oculogyric crisis—involuntary upward conjugate deviation of eyeballs.
2. Skew deviation of eye—one eye is down and medial while the other eye goes up and lateral; seen in acute cerebellar lesion.
3. Ocular bobbing—periodic brisk downward movement of both eyes, and is a feature of pontine lesion. Ocular dipping is brisk upward and slow downward movement of both eyes.
4. Opsoclonus—coarse, irregular, disorganised, chaotic, dancing and non-rhythmic oscillations of eyeball occurring in both vertical and horizontal plane, and persisting for a long period. It is often found in cerebellar lesion, encephalitis or brainstem damage.
5. Ocular flutter—pendular horizontal oscillations on fixing gaze on an object; a feature of cerebellar lesion.

Tests of VIth cranial nerve :

In VIth nerve lesion, the patient usually complains of dizziness or vertigo, tinnitus (ringing in the ears) or deafness.

- (A) Cochlear functions (function of hearing)—first examine the external acoustic canal using a torch and auroscope (wax, tympanic membrane). Hearing should be tested in each ear in turn.
 - a) Speak loudly from a distance of 20 feet, and gradually come closer to the patient. Normal conversational voice should be heard from 20 feet.
 - b) Whisper test—In health, a person is able to hear the whisper from a distance of approximately 10 feet. First whisper from a distance and then gradually move closer to the patient, or test either by the ticking sound of a wrist-watch (watch test) or by rubbing fingers; the other ear should be occluded by finger pressure on the tragus.
 - c) Rinne's test and Weber's test—Read from 'Bedside clinics in Medicine, Part II'.
- (B) Vestibular functions—
 - a) Observe for spontaneous nystagmus—fast phase is away from the side of lesion.
 - b) Positional nystagmus (see above).
 - c) Caloric test—Read from any standard ENT text book.

* Deafness is of two types ;

- I. Conductive deafness—As a result of impacted wax, damage to tympanic membrane, otosclerosis, CSOM etc (i.e., diseases of the external auditory meatus, middle ear and eustachian tube).
- II. Sensorineural deafness—Due to damage of cochlear nerve and organ of Corti. Meniere's disease, acoustic neuroma, drugs like aminoglycosides, fracture of petrous part of temporal bone, pontine lesion etc.

** In health and sensorineural deafness, air conduction is greater than bone conduction.

Case 92

BELL'S PALSY

What is your diagnosis ?

This is a case of right-sided LMN type of facial palsy, probably Bell's palsy.

* Sir Charles Bell (1774-1842) was a Scottish surgeon at Middlesex Hospital, London, UK; Professor of Surgery, Edinburgh afterwards.

Why do you say 'probably' ?

I say 'probably' because I am not allowed to know the history (exposure to cold etc.) and the initial symptoms (pain behind the right ear).

* Facial paralysis may be supranuclear (UMN type), and nuclear/infranuclear (LMN type).

Why do you say so ?

Because the right side of face on inspection shows :

1. Smoothening of furrows in the forehead.
2. Wide palpebral fissure [due to loss of tone of the lower eyelid and unopposed action of the levator palpebrae superioris (LPS)]—lagophthalmos or unequal palpebral fissure in comparison to other side.

3. Asymmetry of blinking.
 4. Abnormality in volitional or reflex movement like smiling or crying.
 5. Watering from the eye or epiphora (due to loss of capillary action as the punctum is no more in contact with conjunctiva).
 6. **Bell's phenomenon** (the eyeball rolls upwards and inwards during attempted forced eye closure. It is a normal as well as protective phenomenon and is preserved in LMN type palsy only).
 7. Loss of nasolabial fold or flattening of the fold.
 8. Angle of the mouth is drawn to left side when the patient is asked to show his upper teeth.
 9. Drooping of the angle of the mouth (right side).
 10. Flapping in and out of the cheek on right side during respiration due to the paralysis of buccinator muscle.
 11. Saliva dribbles from the right angle of the mouth; difficulty in spitting.
 12. Inability to blow or whistle properly (air escapes from the right side of the mouth).
- * An important complaint by the patient is collection of food materials in the affected vestibule during eating. Patient may also complain of numbness and stiffness of the cheek on the affected side though the sensory functions of face remain absolutely intact.
- ** Bell's palsy does not lead to ptosis.
- *** The patient may complain of slurred speech for obvious reasons (i.e., weakness of facial muscles).

Test the individual muscle of face :

Test the muscles on both sides of face and observe the weakness on right side (in this patient).

1. Ask the patient to wrinkle his forehead (either the patient will do it on your command or keeping the head fixed, ask him to look at your index finger which is placed above his head)—Frontal belly of occipitofrontalis (muscle for surprise).
 2. Ask the patient to frown — Corrugator superciliaris.
 3. Ask him to close both eyes forcibly and you try to open the eyelids by your fingers (both eyes must be examined for comparison)—Orbicularis oculi.
 4. Ask him to show his upper teeth, or smile—Levator anguli oris, zygomatic major and minor, depressor anguli oris, buccinator and risorius.
 5. Ask him to blow or whistle, or inflate the mouth with air—Orbicularis oris and buccinator.
 6. Ask the patient to purse his lips shut—Only orbicularis oris.
 7. Ask the patient to retract and depress the angle of the mouth with clenching of teeth (or ask the patient to try to touch the chin to the sternum though he will not move the chin at all from its original position)—Folds of platysma (muscle for grinning) may be seen in the neck.
- * There may be apparent deviation of the tongue to the healthy side on protrusion.
- ** No. 1, 2 and 3 are ideal tests for 'upper face' and No. 4 for 'lower face'.

Table 36 : Differentiation between UMN and LMN type of Vllth nerve palsy :

UMN type	LMN type
<ol style="list-style-type: none"> 1. * 'Upper face' escapes (forntal belly of occipitofrontalis, corrugator superciliaris and upper part of orbicularis oculi escape paralysis) 2. Bell's phenomenon—Never occurs. 3. Facial muscles are not atrophied 4. Taste sensation is preserved 5. Corneal reflex—Preserved 6. Usually associated with same-sided hemiplegia 7. Plantar response on the paralysed side of face—Extensor 	<ol style="list-style-type: none"> 1. Total face is involved on one side 2. Bell's phenomenon—Present 3. There may be fasciculation or muscle atrophy on affected side 4. Taste sensation may be lost 5. Corneal reflex—Lost 6. May be an isolated phenomenon (Bell's palsy) and if associated with hemiplegia, it is always crossed 7. Plantar response on the paralysed side of face—Flexor (may be extensor on the opposite side)

- * i.e., forehead wrinkling, frowning, and eye closure and blinking are unaffected.
- ** In UMN facial palsy—causes weakness of the lower part of face on the opposite side. In LMN facial palsy—causes unilateral complete weakness which is same-sided.
- *** 'Upper face' and 'lower face' are divided by an arbitrary horizontal line passing through outer and inner angles of both the eyes.

What else do you want to examine in your patient of Bell's palsy ?

1. Taste sensation of anterior 2/3rd of the tongue (lost in LMN type).
2. Plantar response (mentioned above).
3. Corneal reflex—Lost in LMN type.
4. Evidence of cerebellar signs—if present on the side of facial palsy, the lesion is probably present at the cerebello-pontine angle (e.g., acoustic neuroma). In cerebello-pontine angle tumour, features of Vth (loss of corneal reflex), VIth and VIIth cranial nerves lesion as well as cerebellar signs along with pyramidal signs are elicited.
5. Hyperacusis.
6. Examination of VIIth cranial nerve (affected in acoustic neuroma).

Examination within the oral cavity in Bell's palsy :

1. Food materials present within the vestibule of mouth on the affected side, and
2. Loss of taste sensation (if nerve to chorda tympani is affected).

What is mimic facial palsy ?

There are three types of facial palsy :

1. UMN palsy,
2. LMN palsy, and
3. Mimic palsy.

Mimic palsy is a rare phenomenon where the emotional movement of the facial muscles is lost but the voluntary movement is preserved i.e., *in other words, facial palsy or asymmetry is visible only on emotional movements e.g., in laughing or weeping.* This condition is seen in frontal lobe lesion because the UMN fibres concerned with emotion have a different course and is separate from the pyramidal tract.

As LMN is the final common pathway, LMN facial palsy affects the voluntary, emotional as well as associated movements.

In UMN facial palsy (obviously with hemiplegia), the paralysed angle of the mouth will be retracted during laughing (emotional movement) and during forcible closure of the eyes (associated movement).
Summary :

- a) UMN facial palsy—No mimic palsy (i.e., emotional movements are preserved).
- b) Frontal lobe lesion—Contralateral mimic palsy (commonly seen in multiple sclerosis).
- c) LMN facial palsy (e.g., Bell's palsy)—All movements lost.

* So, **UMN palsy** includes the classic UMN type paralysis and rare mimic palsy; **LMN palsy** includes both nuclear and infranuclear lesion.

Why the upper face escapes in UMN palsy ?

The upper part of VIIth nerve nucleus (so the upper facial muscles or upper face) is innervated by corticobulbar fibres from both cerebral hemispheres whereas the lower part of VIIth nerve nucleus (so the lower face) receives fibres from opposite hemisphere only. As the upper facial muscles have dual supply, they escape in UMN palsy but LMN is the final common pathway and thus, total face is affected in LMN palsy.

Causes of unilateral LMN type VIIth cranial nerve palsy :

- | | |
|---|-------------------------------------|
| 1. Bell's palsy (commonest and benign). | 6. Sarcoidosis. |
| 2. Leprosy. | 7. Acoustic neuroma. |
| 3. Trauma to parotid gland. | 8. Middle ear disease (e.g., CSOM). |
| 4. Parotid tumour. | 9. Fracture of base of the skull. |
| 5. Herpes zoster infection. | 10. Pontine neoplasm. |

Causes of bilateral facial palsy of LMN type (facial diplegia) :

1. Acute infective polyneuritis (G.B. syndrome).
 2. Leprosy.
 3. Sarcoidosis (Heerfordt syndrome).
 4. Forceps delivery (trauma).
 5. Leukaemia (ALL) or lymphoma.
 6. Bilateral Bell's palsy (rare).
 7. Bilateral otitis media.
 8. Diphtheria.
 9. Lyme disease.
 10. Seroconversion phase of HIV infection.
- * Myasthenia and myopathy produce facial weakness; there is no involvement of facial nerve.

D/D of alteration of facial contour :

1. Hemiatrophy (e.g., localised scleroderma).
2. Hemihypertrophy.
3. Residual Bell's palsy.
4. Lipodystrophy.
5. Unilateral facial oedema (postural).
6. Paget's disease, fibrous dysplasia.
7. Absence of condyle of mandible (congenital)
8. Massive swelling of parotid glands.
9. Acromegaly.
10. Micrognathia.

How do you diagnose bilateral UMN type facial palsy ?

Remember, in bilateral UMN type of lesion, the total face is paralysed (upper 1/2 of face does not escape).

Bilateral UMN type of lesion is diagnosed by (differentiate from bilateral LMN type facial palsy) :

1. Mask like facies.
2. Absence of Bell's phenomenon.
3. Brisk jaw jerk.
4. Presence of glabellar reflex.
5. Spared emotional fibres, and
6. Associated bilateral long tract signs.

* Bilateral UMN type facial palsy is seen in diffuse cerebral atherosclerosis, pseudobulbar palsy and double hemiplegia.

** Isolated unilateral UMN facial palsy is rare but may be seen in CVA and multiple sclerosis.

Definition of Bell's palsy :

It is an acute, idiopathic, unilateral (commonly) LMN type of facial nerve involvement due to non-suppurative inflammation (viral, e.g., often herpes simplex) of the nerve within the facial canal above the stylomastoid foramen.

Inflammation or oedema strangulates the nerve at the stylomastoid foramen and precipitates the paralysis. Exposure to cold or chill predisposes to Bell's palsy (there may be positive H/O travelling by bus or train, sitting by the side of an open window).

Bell's palsy affects all ages and both sexes. There is sudden unilateral facial weakness, which may be associated with loss of taste sensation and hyperacusis. There is no objective sensory loss in face. It is primarily diagnosed on clinical grounds.

How do you assess the progressing lesion in Bell's palsy ?

1. The inflammation may involve the nerve to chorda tympani — There is loss of taste sensation (hypogeusia) in the anterior 2/3rd of the tongue (chorda tympani arises from the facial nerve about 6 mm above the stylomastoid foramen).
2. Nerve to stapedius may be involved — The patient will complain of hyperacusis due to paralysis of stapedius muscle (sound on the paralysed side seems unusually loud).

* These two features indicate a bad prognosis in Bell's palsy.

Syndromes in nuclear type of facial palsy :

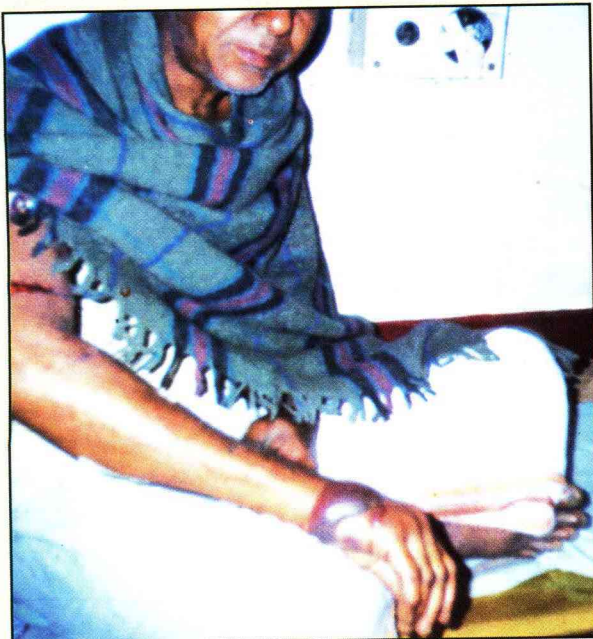
Millard-Gubler syndrome—Paralysis of VIth cranial nerve ± VIIth cranial nerve with contralateral hemiplegia.

Foville's syndrome—Ipsilateral paralysis of conjugate gaze ± VIIth cranial nerve palsy with contralateral hemiplegia.

Clinical features in lesion at the geniculate ganglion :

It is known as **Ramsay Hunt syndrome** which is the affection of geniculate ganglion by herpes zoster (shingles) infection. The clinical features are,

1. Fever with chill and bodyache.
2. LMN type of ipsilateral facial palsy (identical to Bell's palsy).
3. Loss of taste sensation in the anterior 2/3rd of the tongue (on affected side).
4. Hyperacusis (on affected side).



Haemorrhagic bullous lesion in pemphigoid



Vasculitis (Wegener's granulomatosis). Raynaud's phenomenon (colour changes in the tip of the fingers) with destruction of nose and nasal septum is seen. Hands are very cold on palpation



Facial hematoma, mild periorbital hematoma (raccoon eyes) and blood collection in forehead in a case of head injury



Multiple discharging fistula in lower abdomen in Crohn's disease



Loss of elasticity of skin in severe dehydration – 'the sign of ridge', usually tested over the sternal area



Amyloidosis – Clustured waxy papules in the right side of the neck and periorbital ecchymoses ('raccoon eyes'). She also had macroglossia



Cachexia (anaemia plus emaciation) with sarcopenia (decrease in muscle mass) – all skin and bone condition



Herpes zoster ophthalmicus of right side. There is risk of corneal scarring and secondary panophthalmitis



Formation of **cold abscess** over the sternal area. Scars of healed tuberculous lymphadenitis in the right axilla and neck are also noted



Molluscum contagiosum – multiple, glistening pale, poxvirus-induced, umbilicated, dome-shaped, pinhead to 1 cm papules. The child is HIV positive

- Herpetic rashes on tympanic membrane, external auditory canal, pinna, and occasionally over pharynx, soft palate and tongue.

How do you elicit corneal reflex ?

Method — the patient is asked to see into the distance or at the ceiling. The cornea at its conjunctival margin is touched lightly with a damp cotton wool which is twisted into a fine hair. If the reflex is present, there is simultaneous prompt closure of both the eyes. Both the eyes should be tested one by one. The cornea is stimulated from the side to avoid 'menace reflex' (reflex closure of the eyes, if an object is brought to the patient directly from the front).

Afferent path — Vth cranial nerve (ophthalmic division).

Efferent path — VIIth cranial nerve.

Centre — Pons.

Response — Closure of both the eyes.

* Never touch the central part of cornea because corneal ulceration may develop in the presence of corneal anaesthesia. In the absence of cotton, blowing a puff of air into each cornea will serve the purpose. This reflex is also known as *conjunctival reflex*. Loss of corneal reflex is an early sign of sensory involvement of trigeminal nerve in cerebello-pontine angle tumour.

** While eliciting the corneal reflex, prompt closure of ipsilateral eye (eye being examined) is known as direct corneal reflex while closure of contralateral eye is known as consensual corneal reflex.

Management of Bell's palsy :

- Prednisolone**—A course beginning with 60-80 mg daily, orally during the first 5 days and then tapered over the next 5 days may be of some help; ideally started within 72 hours.
- Aspirin or other **NSAID** (non-steroidal anti-inflammatory drugs) may be given for relief of pain and inflammation.
- Eye padding is done during sleep and local antibiotic drop is instilled in the eye, if signs of congestion is seen.
- Proper mouth wash is advised after each meal.
- Gentle massage over the paralysed muscles is advised.
- Splint is used to prevent drooping of the lower part of face (by leukoplast).
- Facial exercise is advised (standing in front of a mirror) or **consult physiotherapist**.
- Galvanic current stimulation of the paralysed muscles may be of some help.
- If not improved at all within 6 weeks — Surgical decompression may be done at the stylomastoid foramen (majority of clinicians say that the indications for operation are still not clear). Suturing upper and lower eyelid (tarsorrhaphy) may be done if eye closure is not at all possible.
- Parenteral vitamin B₁₂, B₆ and B₁ may be given; oral or parenteral methylcobalamin may be of some help.
- Recently, acyclovir (400 mg five times a day for 10 days) or valacyclovir (1000 mg daily for 5-7 days) is tried, although the evidence for giving antivirals is poor.

* Bell's palsy usually recover (70-80%) spontaneously within 2-12 weeks; < 10% may have residual weakness.

Features of incomplete recovery from VIIth nerve palsy :

- Residual paralysis on the affected side of face.
- Hemifacial spasm (irregular, painless clonic spasm of facial muscles).
- Crocodile tears (unilateral lacrimation during chewing foods) — Rare.
- Unwanted facial movements (e.g., closure of eyes with movement of mouth).

Complications of Bell's Palsy :

- Exposure keratitis or conjunctivitis.
- Hemifacial spasm.
- Crocodile tears.
- Facial contracture.
- Jaw winking.
- Social embarrassment.

Bad prognostic factors :

- Complete palsy at the beginning.
- Associated co-morbidities, e.g., hypertension or diabetes.
- Hyperacusis or loss of taste sensation.
- Severe axonal degeneration on electrophysiological study (EMG) after 10 days.

Identification points :

Observe the face for a) asymmetry, b) symmetry of blinking and eye closure, and c) spontaneous

movements of face. Lastly, ask the patient to show his upper teeth when the angle of the mouth is drawn to the healthy side. Try to test the taste sensation, if possible.

* Read the examination of taste sensation from the chapter on 'Examination of the tongue'.

** Facial nerve supplies all muscles of the face and scalp, except levator palpebrae superioris. In the parotid region, facial nerve divides into temporofacial (temporal, zygomatic and upper buccal) and cervicofacial (lower buccal, mandibular and cervical) branches to supply muscles of face, scalp and platysma.

*** Recurrent facial palsy may be a feature of multiple sclerosis.

Case 93

CHOREA

What is chorea ?

These are brief, rapid, jerky, explosive, non-repetitive, quasi-purposive involuntary movements chiefly involving the face, tongue and limbs. The movements are totally irregular in time, rhythm, character and place of occurrence. Chorea literally means 'a dance'.

Types of chorea :

1. Sydenham's chorea or Saint Vitus' dance (rheumatic).
2. Huntington's chorea (autosomal dominant disorder of early middle life with dementia).
3. Chorea gravidarum (during pregnancy).
4. Senile chorea (follow vascular lesion in old age; mainly affects oral-buccal-lingual muscles).
5. Chorea mollis (with hemiplegia).

Other aetiology :

- | | |
|--|---|
| 6. OC pills, L-dopa, phenytoin, phenothiazines. | 11. Chronic liver disease (e.g., Wilson's disease). |
| 7. Systemic lupus erythematosus and antiphospholipid syndrome. | 12. Alcohol. |
| 8. Polycythemia vera. | 13. Hypernatraemia. |
| 9. Thyrotoxicosis. | 14. Lacunar infarction. |
| 10. Post-encephalitic. | 15. Hypoparathyroidism. |
| | 16. Carbon monoxide poisoning. |

How to demonstrate chorea ?

The diagnosis of chorea is made on clinical grounds. Ask the patient to raise up the hands over his shoulders and to maintain the posture. If chorea is present, patient will start rapid as well as jerky movements of the upper limbs.

* Very often agitation or excitement (e.g., a sudden slap over patient's face) increases chorea; it disappears during sleep.

Six cardinal features in chorea :

1. **Hypotonia** (though an extrapyramidal disorder) — Assessed by decreased resistance to passive movement. It is the only extrapyramidal lesion having hypotonia.
2. Patient will tend to pronate the outstretched hands (and sway outward) while extending the arms above the head — The '**pronator sign**'.
3. Ask the patient to grasp or squeeze the examiner's hand or finger tightly. There is waxing and waning of the grip — This is known as "**milkmaid's grip**" or '**milking sign**', and is due to irregular contraction of fingers.
4. The patient is asked to stretch out the hand and spread the fingers — Elbow is hyperextended, forearm is hyperpronated, wrist joint is flexed, metacarpophalangeal joints are extended with the separation of fingers (dinner-fork deformity). This is a manifestation of hypotonia. This phenomenon will be more obvious, if the patient is asked to raise up the hands above his head. This is also known as '**spooning sign**'.
5. Knee jerk — Pendular (due to hypotonia) and often the jerk is called '**hung-up reflex**' (when a choreic movement is superimposed on a tendon reflex).

6. Ask the patient to protrude the tongue — The patient protrudes it momentarily and takes it back within the oral cavity instantaneously (**lizard tongue**). The tongue, when projected, looks like a 'bag of worms'. Typical grimacing of face with protrusion of tongue often clinches the diagnosis.

* Clinical triad of chorea are : hypotonia, choreiform movement and mental instability.

What is athetosis and tics ?

Athetosis — These are slow, sinuous, purposeless, writhing involuntary movements usually involving the distal part of a limb (athetosis means 'instability of posture'). The movements are best seen in fingers, hands, toes, feet and tongue (snake charmer's movement).

Tics — Simple, normal movements which are repeated unnecessarily to become a social embarrassment. These are 'stereotyped' movements like head nodding, eye blinking or grimacing of face. Tics may be seen in anxiety states. They can be briefly suppressed by the patient.

Aetiology of **athetosis** are :

- | | |
|--|--------------------------------|
| 1. Post-hemiplegic (infantile). | 5. Birth injury. |
| 2. Wilson's disease. | 6. Encephalitis. |
| 3. Kernicterus. | 7. Cerebral palsy. |
| 4. Overdose of L-dopa, phenothiazines. | 8. Degenerative brain disease. |

Table 37 : Differentiation between chorea and athetosis

Features	Chorea	Athetosis
1. Lesion	1. Caudate nucleus	1. Putamen
2. Tone	2. Hypotonia	2. Hypertonia (rigidity).
3. Basic pattern	3. Rapid, continually interrupted and never proceeding to completion	3. Slow movements; extension and pronation plus flexion and supination of the arm, with alternating flexion and extension of fingers
4. Effect of agitation or excitement	4. Often increased	4. No effect
5. Parts involved	5. Proximal > distal part	5. Usually distal > proximal
6. Tongue	6. Lizard tongue	6. Chewing tongue
7. Deep reflexes	7. Pendular or hung-up reflex	7. Normal

What is 'pure chorea' ?

Chorea is a delayed manifestation of rheumatic fever (Jones major criteria) and other manifestations may not be present when chorea appears. Arthritis always subsides before the appearance of chorea. Sydenham's chorea appears after a long latent period (upto several months) from the antecedent streptococcal infection. When no previous rheumatic manifestations are observed, the cases are known as 'pure chorea'.

* Hemichorea—choreic movements restricted to one half of the body. Usually chorea is bilateral.

Drugs used in the treatment of chorea :

- | | |
|--------------------|----------------------|
| 1. Phenobarbitone. | 4. Haloperidol. |
| 2. Clonazepam. | 5. Sodium valproate. |
| 3. Chlorpromazine. | 6. Tetrabenazine. |

What are the differential diagnosis of chorea ?

1. Athetosis.
2. Dyskinesia (mainly affects the pharyngeal, perioral and neck muscles, and produces axial tortional movements. This movement disorder is mainly drug-induced e.g.. by metoclopramide, phenothiazines, levodopa etc).
3. Tics.
4. Hemiballismus (violent ballistic and flinging movement of one side of body).
5. Myoclonus (sudden, rapid, irregular, jerky involuntary movements of a limb due to contraction of a single muscle or a group of muscles).

Effect of sleep on chorea :

Choreic movements subside.

What are the other involuntary movements you know ?

Other than, 1. Chorea. 2. Athetosis, and 3. Tics (habit spasm),

There are,

- | | |
|--|--|
| 4. Myoclonus, | 9. Hemiballismus, |
| 5. Convulsions, | 10. Spasmodic torticollis. |
| 6. Tremor, | 11. Myokymia (called 'false or benign fasciculation'), |
| 7. Dyskinesia, | 12. Asterixis, |
| 8. Dystonia (abnormally maintained posture). | 13. Tetany (see the section on 'Fasciculation'), and |
| | 14. Muscle cramp. |

* Fasciculation, titubation and opisthotonus are involuntary movements too. Tremor is a very common involuntary movement in clinical practice. Few clinicians do not agree that convulsions and muscle cramp are involuntary movements.

** Akathesia is motor restlessness with inability to sit still. There is always an intense urge to move. It is commonly seen in parkinsonism, chorea, thyrotoxicosis or phenothiazine therapy.

*** Restless leg syndrome (Ekbom syndrome)—intense desire to move the legs due to unpleasant sensation there, specially when the patient attempts to sleep or sits quiet. This is seen in chronic renal failure, peripheral neuropathy and iron deficiency anaemia.

Chorea without hypotonia :

Probably, it is a patient of chorea associated with athetosis (choreo-athetosis).

Conclusion :

1. Classical movements are already described. When choreic movements are superimposed on voluntary movements, there may be a 'grotesque' (strange or ugly) look of the posture.
2. Observe the face and movements of the tongue. At rest, the patient is 'unable to sit still'.
3. Inability to perform a voluntary movement in the absence of paralysis.
4. Bizarre gait.
5. Demonstrate '6 cardinal features'.

* Always examine the heart for any valvular lesion; search for exophthalmos, butterfly rash in face, K-F ring in cornea or polycythemia.

Case 94**FASCICULATION****What is fasciculation ?**

These are fine, rapid, irregular and inconstant visible twitching movements of large group of muscle fibres or the whole motor unit. **It indicates LMN lesion.**

What is fibrillation ?

These are contractions of a single muscle fibre. It is a state of hyperirritability (denervation hypersensitivity) of single muscle fibre and usually can not be detected in naked eye. It can be diagnosed by electromyography (EMG) and usually signifies complete denervation of individual muscle fibre.

Methods of bedside demonstration of fasciculation :

1. They may be seen as ripples underneath the skin overlying muscles at rest or sometimes 'spontaneously' produce clinically visible contractions.
2. If fasciculation is not spontaneous, they may be evoked by mechanical stimulation. In the presence of proper light, sharply tap (by the conical end of a hammer) over the degenerating muscle (never on fully degenerated muscle) for demonstration of fasciculation. Usually the bigger muscles like biceps, triceps, deltoid, calf muscles are chosen (this procedure is controversial because fasciculation should be spontaneous).

3. Fasciculation of the tongue — Keep the tongue relaxed in the floor of the mouth (never protrude the tongue) for demonstration of fasciculation.

Significance of fasciculation :

1. It signifies chronic degeneration of anterior horn cells or their homologue cranial nerve nuclei. It is always seen in a muscle which has undergone wasting. In the absence of wasting, fasciculation has no significance (benign fasciculation). So **fasciculation is seen in muscles undergoing active degeneration, not totally degenerated yet (wasting but not wasted).**
2. If fasciculation disappears from a muscle (which showed fasciculation previously), it signifies that the muscle is totally degenerated i.e., the anterior horn cells are completely destroyed (prognostically worse).
3. The muscular contraction of this special type of involuntary movement (fasciculation) is of fine amplitude and thereby, it is not able to move the adjoining joint.

* Fasciculation is due to spontaneous firing of surviving axons to innervate the denervated muscle fibres.

Can you see fibrillation in naked eye ?

As previously discussed, it is diagnosed by EMG but sometimes it is clinically evident in tongue as tongue has no submucous coat.

Common causes of fasciculation :

- | | |
|---|--|
| 1. Chronic motor neurone disease (MND). | 6. Intramedullary tumour. |
| 2. Cervical spondylosis. | 7. Peripheral neuropathy. |
| 3. Syringomyelia or syringobulbia. | 8. Acute stage of poliomyelitis. |
| 4. Peroneal muscular atrophy. | 9. Hypoxia, hypercapnoea, hypoglycaemia. |
| 5. Recovery from facial palsy (Bell's palsy). | 10. Organophosphorus poisoning. |

* MND (amyotrophic lateral sclerosis) is the commonest cause. Fasciculation may be evoked by voluntary muscular contraction, cooling the muscle by ethyl chloride spray or hyperventilation.

What is benign fasciculation ?

When fasciculation is present in the absence of muscular wasting, it is known as 'benign fasciculation'. It is commonly seen in anxiety and fatigue states. It is often evident in thenar eminences, thighs, around the shoulder joint or orbicularis oculi (myokymia) muscles.

How do you classify chronic motor neurone disease (MND) ?

This is a disease of unknown origin which leads to degeneration of anterior horn cells of the spinal cord, motor nuclei of cranial nerves, and the corticospinal and corticobulbar pathways. The disease is characterised by insidious onset with a steadily progressive course, in a middle aged or elderly person. The classification goes like :

- (A) Progressive muscular atrophy (predominant LMN type)—Weakness, wasting and fasciculation.
- (B) Amyotrophic lateral sclerosis (predominant UMN type in lower limbs, and combined LMN plus UMN type in upper limbs) — Commonest (see below).
- (C) Primary lateral sclerosis (predominant UMN type)—Progressive quadriparesis (rare).
- (D) Progressive bulbar palsy.
- (E) Pseudobulbar palsy.
- (F) Mixed type of D and E.

* **MND may be given as a long case.**

Describe the motor functions' in amyotrophic lateral sclerosis (MND) :

(A) Nutrition of muscles :

1. **Wasting of the small muscles of hands (skeleton hand)**, prominence of long flexor and extensor tendons of hands; sometimes, there is presence of claw hand. Knuckles are prominent and interosseous spaces are depressed.
2. At first there is wasting of the small muscles of hands which is followed by involvement of forearm muscles. Lastly, there is wasting of deltoid, supraspinatus etc.
3. There is not much wasting of lower limbs.

(B) Tone of muscles :

- Upper limb — Tone is usually decreased (hypotonia).
Lower limb — Increased (clasp-knife spasticity).

(C) Power of muscles :

Power Is decreased more In upper limb than in lower limb because of the combined UMN and LMN involvement.

(D) Involuntary movements :

Fasciculation is seen predominantly in the upper limbs. Presence of fasciculation helps in the clinical diagnosis of MND. Never forget to see the tongue for fasciculation.

(E) Coordination of movements : Normal in both limbs (severe wasting may hamper coordination).

* There is early weakness of respiratory muscle. Usually there is no bed sore formation due to preserved sensory function.

** Rationality of nomenclature : LMN involvement—muscle atrophy (i.e., amyotrophy), UMN involvement—loss of fibres of lateral column (fibrillary gliosis i.e., lateral sclerosis).

*** The course of the disease is spastic quadriparesis or paraparesis with wasting.

Reflexes in MND :

(A) Superficial reflex :

- (i) Abdominal reflexes are lost. It is seen that abdominal reflex may not be lost for a considerable period in MND though there is bilateral UMN lesion.
- (ii) Cremasteric reflex — Lost.
- (iii) Plantar response — Bilaterally extensor.

(B) Deep reflexes :

- (i) Upper limb Jerks are brisk in the presence of wasting (clinically very characteristic of amyotrophic lateral sclerosis).
- (ii) Lower limb — Jerks are brisk, clonus may be seen.

(C) Visceral reflexes : Normal.

N.B. . Small muscles of hands, forearm, deltoid, tongue, facial muscles are involved early. *Higher functions, external ocular muscles, autonomic, cerebellar, extrapyramidal, sensory and sphincter functions are normal in MND.* So, the disease presents with spasticity, brisk jerks, Babinski's sign in the lower limb, and fasciculation, weakness, wasting in the upper limb. Electromyography (EMG) is diagnostic (shows chronic partial denervation); nerve conduction velocity (NCV) is usually normal. Progressive and fatal course within 3-5 years. Riluzole (100 mg/day) may slow progression.

UMN Palsy in lower limb and LMN palsy in upper limb :

1. Amyotrophic lateral sclerosis.
2. Cervical spondylosis.
3. Syringomyelia.
4. Friedreich's ataxia.
5. Cervical extramedullary compression.
6. Patchymeningitis hypertrophic cervicalis (spinal syphilis).

What is tetany ?

Tetany is increased neuromuscular irritability due to low ionic calcium in blood. Normal serum calcium level is 9-11 mg/dl and the ionic fraction is 4.5-5.6 mg/dl.

Serum calcium < 8.5 mg/dl is hypocalcaemia while >11 mg/dl is regarded as hypercalcaemia.

Classical physical signs in tetany :

The increased neuromuscular irritability is tested by :

1. **Trousseau's sign** - When the pressure is raised above the systolic BP for 3 minutes, typical carpal spasm appears. There is flexion of MCP joints with extension of interphalangeal joints and the flexed thumb takes its position in between the index and the middle fingers (opposition of thumb). This is known as "main d' accoucheur" or obstetrician's hand. Pedal spasm is less frequently demonstrated. Latent tetany is best recognised by eliciting this sign.
2. **Chvostek's sign** - Tapping the facial nerve (by finger or hammer) in front of the ear will produce twitching of facial muscles.
3. **Erb's sign** - Muscular contractions can be produced by application of subthreshold electrical stimulation (0.5-2.0 milli-amps).

4. **Peroneal sign** - Tapping the peroneal nerve at the neck of fibula will produce plantiflexion and adduction of the foot with extension of knee (pedal spasm).
5. IECG — Prolonged Q-T interval.]

N.B. : Symptoms of tetany are.

1. Irritability.
2. Muscle cramps (carpopedal spasm).
3. Peripheral (hand and feet) and circumoral paraesthesia.
4. Triad of symptoms are common in children i.e., carpopedal spasm, laryngismus stridulus (stridor, respiratory distress, cyanosis), and convulsions.
5. Dysphagia, dyspnoea, dysuria, abdominal colic etc.

Common causes of hypocalcaemia :

1. Malabsorption syndrome.
2. Hypoparathyroidism or pseudohypoparathyroidism.
3. Acute pancreatitis.
4. Chronic renal failure (CRF).
5. Rickets and osteomalacia.
6. Hypoalbuminaemia.
7. Alkalosis due to hyperventilation (often hysterical) or persistent vomiting (pyloric stenosis).
8. Citrated blood in massive transfusion.
9. Drugs—Calcitonin, bisphosphonates.

* Low total calcium is not associated with tetany if ionised fraction is normal. Low ionised calcium is associated with tetany irrespective of total calcium level. Transient hypocalcaemia (burn, sepsis) does not develop into tetany.

** Tetany is rare in chronic renal failure because of the presence of metabolic acidosis.

*** Causes of **hypercalcaemia** are : Primary hyperparathyroidism, malignant tumours, prolonged immobilisation, ingestion of excess vitamin A or D, milk-alkali syndrome, Paget's disease, thiazide diuretic, Addison's disease, granulomatous disease (e.g., sarcoidosis).

Causes of lock jaw or trismus :

Trismus develops due to sustained involuntary spasm of masseter and temporalis (masticatory muscles) resulting in inability to open the mouth and mastication, and is found in :

1. Tetanus (spatula test is positive; trismus is by far the most important early symptom)*.
2. Impacted wisdom tooth.
3. Peritonsillar abscess (quinsy), dental abscess, Ludwig's angina.
4. Acute follicular tonsillitis.
5. Temporo-mandibular osteoarthritis (Costen's syndrome) or rheumatoid arthritis.
6. Sometimes in drug-induced dyskinesia (metoclopramide, phenothiazines).
7. Sometimes in tetany, strychnine poisoning, cyanide toxicity.
8. Parotitis, mumps.
9. Hydrophidae group of snake bite.
10. Rabies.
11. Anaesthesia-induced malignant hyperthermia.
12. Hysterical.

N.B. : Painful trismus—2, 3, 4; painless trismus—tetanus and others.

* **Spatula test**—In health, touching the posterior pharyngeal wall by spatula produces reflex opening of mouth. In tetanus, paradoxically mouth closes in such a way that the spatula can not be taken out easily. Thus, spatula test is positive in tetanus and negative in others.

** In scleroderma and submucosal fibrosis of oral cavity, the patient may find difficulty in opening the mouth; but this is not true trismus.

*** **'Facies' in tetanus**—characteristic risus sardonicus (grimace mimicking 'smile of satan') due to contraction of facial muscles, and blepharospasm.

Case 95

MYOTONIA

What is myotonia :

In certain muscle disease like dystrophia myotonica (an autosomal dominant disorder), the relaxation of the muscle is impaired following a strong contraction (*myotonia*). This continued, involuntary muscular contraction after cessation of voluntary effort is known as myotonia. Myotonias are also classified as 'channelopathies' as there is defective skeletal muscles cell chloride ion membrane conductance.

Types of dystrophies of the muscle :

There are two types mainly :

1. Muscular dystrophy (Duchenne type etc.) or myopathy, and
2. Myotonic dystrophy (or dystrophia myotonica).

* The two most common myotonias are myotonic dystrophy and myotonia congenita.

How do you diagnose myotonia at the bedside ?

The phenomenon (described below) is most evident when the muscles are cold. Myotonic muscle shows sustained contraction on direct percussion. The different bedside methods are :

1. Shake hand with the patient and then let it go suddenly — The patient's grasp is maintained for a moment, then released slowly and gradually i.e., there is poor hand grip relaxation.
2. Percuss the thenar eminence by a hammer — It shows a peculiar movement of the thumb (opponence movement) and a dimple of contraction appears on thenar eminence which fills up very slowly ('percussion myotonia'); this can also be tested over the wrist extensors.
3. Percuss over the tongue — A dimple of contraction appears which relaxes very slowly.
4. Ask the patient to close the eyes forcibly and then to open the eyes. The patient can not open the eyes promptly or opens it very slowly.
5. Ask the patient to smile and then to stop it — Smile remains fixed for a longer duration.

Clinical features of myotonia dystrophica :

Males are affected more than females.

1. Myotonia.
2. Ptosis (partial) with ophthalmoplegia.
3. Proximal muscle wasting with distal limb muscles weakness.
4. Long facial structure ('hatchet faced' and 'swan neck' appearances are due to atrophy of temporalis/masseter/facial muscles, and sternomastoid muscle respectively); a weak and expressionless face.
5. Cataracts.
6. Frontal baldness.
7. Mental retardation (mild).
8. Testicular atrophy.
9. Alopecia.
10. Dysarthria due to weak palatal, pharyngeal and tongue muscles.
11. Cardiac conduction defects, arrhythmias, mitral valve prolapse, glucose intolerance, insulin resistance, decreased oesophageal and colonic motility, low serum IgG, poor tolerance to general anaesthetics.

* Diagnosis is largely clinical. Serum CK level may be normal or mildly elevated, EMG is diagnostic and shows evidence of myotonia (waxing and waning of potentials, known as *dive-Bomber-effect*), while muscle biopsy shows type I fibres with increased number of nuclei, ring fibres and sarcolemmal masses.

Drugs used in the treatment of myotonia :

1. Phenytoin,
2. Mexiletine,
3. Quinine, and
4. Procainamide.

How to test the temporalis muscle ?

First inspect the suprazygomatic region for the muscle bulk of **temporalis**. In weakness of temporalis, the temporal fossa will be hollow. Now ask the patient to clench his teeth. Inspect (above the zygoma) and palpate the temporalis muscle on both sides. Paralysed muscles will be less prominent and active muscle will stand out.

Masseter, another muscle supplied by the Vth cranial nerve, is tested in the same way. Paralysis of masseter results in flattening of the angle of the jaw. Again ask the patient to clench his teeth. Look for the prominence of masseter and palpate it, which stands out as corrugated hard mass at the angle of the jaw. Masseter, and temporalis to a lesser extent elevate the jaw (jaw closure).

* The other muscles supplied by the Vth cranial nerve are lateral and medial **pterygoids**. Individually, lateral or medial pterygoids help in jaw opening and its side to side movement. When both the pterygoids act, they help in jaw opening and its protrusion. They are tested by asking the patient to depress the jaw and to move it from side to side; then do the test against resistance offered by you by placing your hand under the chin and sides of the chin from time to time. In unilateral pterygoid palsy, on attempted jaw opening, the jaw will deviate (appraised by noting the relation between upper and lower incisor teeth) towards the paralysed side being pushed by the healthy pterygoids (as geniohyoid muscle in tongue).

** Jaw opening is done by pterygoids, mylohyoid and anterior belly of digastric muscle.

How do you test the sternomastoids ?

1. Both sternomastoids - Ask the patient to press the chin downwards against the examiner's resistance. Both sternomastoids will become prominent which can be corroborated both by inspection and palpation of the muscles.
2. Individual sternomastoid - To test the right sternomastoid, ask the patient to turn the chin to left side against resistance by placing your hand on left side of patient's face. The sternomastoid present on the right side becomes prominent on chin movement, if healthy, compare the test with the other side.

* In bilateral sternomastoid weakness, head will drop backwards. 1\vo muscles which are supplied by the XIth cranial nerve are trapezius and sternomastoid. Test the trapezius by asking the patient to shrug his shoulders, against your resistance (first demonstrate the "shrugging" to the patient and then press both shoulders downwards simultaneously from BEHIND).

** Accessory nerve (XIth cranial nerve) has two parts : Cranial (arising from the caudal part of nucleus ambiguus) and spinal (arising from upper five cervical segments). While examining accessory nerve, first observe for wasting and fasciculation of sternomastoid and trapezius, and then examine for drooping of the shoulder. Lastly, test the powers of sternomastoid and trapezius.

Neuroanatomy of the trigeminal nerve :

It is the largest cranial nerve.

Sensation of face is provided through three branches of trigeminal nerves, e.g..

- (i) Ophthalmic branch (V_1)—supplies conjunctiva, cornea, medial part of the skin of the nose as far as the tip, upper eyelids, forehead and scalp as far as the vertex.
- (ii) Maxillary nerve (V_2)—supplies lower eyelids, upper cheek, temple, sides of nose, upper lip, gum/teeth/palate of upper jaw.
- (iii) Mandibular division (V_3)—supplies the ear, lower part of face (except the angle of mandible), lower lip, lower teeth and the epithelium of the anterior 2/3rd of the tongue.

The motor branch supplies the '**muscles of mastication**' i.e., masseter, temporalis, lateral and medial pterygoids. It also supplies anterior belly of digastric, mylohyoid, tensor tympani and tensor

veli palati.

The nucleus of trigeminal nerve are arranged like :

- (i) Principal nucleus (motor and sensory) at pons—the motor branch conveys through V_3 to innervate muscles of mastication; sensory nucleus is meant for touch, joint-position sense).
- (ii) Mesencephalic nucleus—carries unconscious proprioceptive information from palate, temporomandibular joint and muscles of mastication e.g., governs how much pressure is given to break a chicken bone during mastication.
- (iii) Nucleus of spinal tract of trigeminal nerve extends from pons to C_2 level of spinal cord, and is meant for pain and temperature sensation of face.

Tests of Vth cranial nerve :

8

1. Sensory functions Test for light touch, pain and temperature sensation (compare the right with the left).
 - a) Ophthalmic division :
 - (i) On the both sides of root of the nose and forehead.
 - (ii) Test for corneal reflex.
 - b) Maxillary division :

On the both malar region and upper lip.
 - c) Mandibular division :

On the both sides of chin (always avoid the angle of mandible which is supplied by C segment of the spinal cord by great auricular nerve).
2. Motor functions—Test the power of temporalis, masseter and pterygoids (i.e. muscles of mastication).
3. Reflex—Demonstrate the jaw jerk.

2

Clinical pearls in trigeminal nerve :

1. Bruxism involuntary and forceful teeth grinding during sleep.
2. Trismus—sustained involuntary spasm of masseter and temporalis, commonly found in tetanus.
3. Jaw tremor—in parkinsonism.
4. Complex partial seizure—chewing and masticatory movement of lower jaw.
5. Jaw claudication—giant cell arteritis.
6. Jaw fatiguability—myasthenia gravis.

Case 96**MYOPATHY****What is muscular dystrophy or myopathy ?**

These are genetically determined abnormality characterised by progressive degeneration of group
volunta[^]muscles¹ involvement of the nervous system (generally, myopathy describes diseases of

Classification of muscular dystrophy (hereditary myopathy) :

- I. X-linked :
 - a) Severe form of muscular dystrophy (Duchenne type).
 - b) Milder or benign form of X-linked disease (Becker type).
- II. Autosomal recessive :
 - a) Limb-girdle type (usually scapulo-humeral, rarely pelvi-femoral).
 - b) Congenital variety.
- III. Autosomal dominant :
 - a) Facio—scapulo-humeral (Landouzy-Dejerine type).
 - b) Distal myopathy (Gower type).
 - c) Ocular or oculo-pharyngeal.

Why do you say Duchenne type of myopathy (DMD) ?

1. Male patient (due to X-linked inheritance, females carry the disease and males are the victims) — Starts in infancy and childhood with H/O frequent fall, rare after puberty
2. Positive family history (as it is X-linked disease, oblique transmission is present and maternal uncle, uncle or nephew are affected) - If you are allowed to take the history.
3. Insidious onset with gradually progressive course.
4. Mental retardation (may be present).
5. Positive Gowers' sign.
6. Difficulty in climbing upstairs and getting up from sitting position (due to involvement of

pelvi-femoral muscles i.e., **proximal muscles**): pelvic muscles involvement > shoulder muscles.

7. Pseudohypertrophy in few muscles and atrophy in others.

8. **Sensory system, bladder function, reflexes (superficial and deep) - All are normal.**
There is no fasciculation (to differentiate it from MND).

9. Waddling gait.

* Deep reflexes are normal till the late stage.

** At 8-10 years, walking requires use of braces; by the age of 12 years, the patient is wheel chair-bound.

Classification of muscle diseases :

(A) Acquired :

1. Inflammatory myopathies—polymyositis, dermatomyositis, inclusion body myositis.
2. Endocrine and toxic myopathies—Cushing's syndrome, thyroid disease (both hyper- and hypothyroid), hypokalaemia, hypocalcaemia, alcohol abuse, steroids/fibrates.
3. Myasthenic—myasthenia gravis, Lambert-Eaton myasthenic-myopathic syndrome.

(B) Genetic :

1. Dystrophic—see the classification of muscular dystrophy (vide page 472).
2. Myotonic—myotonic dystrophy, myotonia congenita.
3. Channelopathies—hypokalaemic/hyperkalaemic/normokalaemic periodic paralysis.
4. Metabolic myopathies—myophosphorylase deficiency (McArdle's syndrome), mitochondrial diseases, malignant hyperpyrexia.

What are the X-linked disorders ?

- | | |
|----------------------------------|-----------------------------------|
| 1. Haemophilia. | 6. Colour blindness. |
| 2. G ₆ PD deficiency. | 7. Hunter's syndrome. |
| 3. Christmas disease. | 8. Hypophosphataemic rickets. |
| 4. Duchenne muscular dystrophy. | 9. Nephrogenic diabetes insipidus |
| 5. Ocular albinism. | 10. Lesch-Nyhan syndrome. |

* **Y-linked single gene disorder** is manifested commonly by 'hairy ears' in male. Other Y-linked disorders are XY female with the Y chromosome lacking the SRY (sex-region determining Y factor) gene or XY male having the SRY gene.

Pseudohypertrophy versus true hypertrophy :

(A) Pseudohypertrophy :

These are the large muscles as a result of deposition of fibrofatty tissue. The muscles look bulkier than normal muscles and they are doughy in feel (elastic feel is lost). When asked to contract, they are not hard and globular. The muscles are strikingly weak in spite of their large size. They are hypotonic and the deep reflexes may be dull (reflexes involving the muscles). Muscles affected by pseudohypertrophy in DMD are :

1. Calf muscles.
2. Glutei,
3. Quadriceps.
4. Deltoid,
5. Infraspinatus.

Atrophy observed in DMD is classically seen in :

1. Pectoralis major (parchment paper-like),
2. Latissimus dorsi,
3. Biceps.

* Pseudohypertrophy of muscles is seen in DMD, hypothyroidism (percussion myoedema is characteristic), glycogen storage disease and rarely in trichinosis.

(B) True hypertrophy :

Large muscles, elastic in feel; when asked to contract, they are hard and globular. These muscles are stronger than normal-sized muscles.

N.B. : In DMD, usually there is symmetrical involvement of muscles. Ultimately all the muscles in the body become atrophied. Facial muscles and small muscles of the hands are usually spared in DMD.

Causes of true hypertrophy of muscles :

1. Manual labourers.
2. Athletes.
3. Myotonia.
4. Sometimes, in cysticercosis (in muscle).
5. Acromegaly.
6. Benign hypertrophy of limb muscles (hypertrophica musculorum vera of Oppenheim).

What is Gowers' rising test ?

When the patient is asked to stand from lying down position, he adopts a curious manner of rising. He first rolls over his body and then take support successively on the ground, feet, legs, knees and waist, and finally stands up. It seems that the patient is 'climbing up the legs' and is known as Gowers' rising test or Gowers' sign. This sign is not specific for DMD, as it may be found in any condition where the pelvic girdle muscles are weakened and thus, it may be observed in polymyositis, spinal muscular atrophy and other myopathies (e.g., endocrine myopathy).

What is waddling gait ?

It is like the 'gait of a duck'. The body is first tilted backwards, with an increase of lumbar lordosis. The feet are placed wide apart and the body sways from side to side as soon as the patient starts stepping. The hip tilts down when the leg is being lifted. The heel and toes are brought down simultaneously. It is due to weakness of the proximal pelvic muscles and is specially seen in myopathy. Causes of waddling gait are :

1. Duchenne muscular dystrophy.
2. Pregnancy (advanced).
3. Congenital dislocation of hip (bilateral).
4. Sometimes in huge ascites or morbid obesity.

Causes of proximal muscle weakness :

1. Muscular dystrophy.
2. Polymyositis and dermatomyositis.
3. Diabetic amyotrophy.
4. G. B. syndrome.
5. Neuralgic amyotrophy.
6. Radicular lesion, e.g., cervical spondylosis.
5. Charcot-Marie-Tooth disease.
7. Sometimes in fulminant myasthenia gravis.
6. Inclusion body myositis.
8. Thyrotoxic myopathy or Cushing's syndrome.
9. Metabolic myopathy e.g., hypokalaemia.
10. Drug-induced—Corticosteroid myopathy.
11. Malignancy.
12. Osteomalacia
13. Porphyric neuropathy.

Causes of distal muscle weakness :

1. Wasting of the small muscles of hands
(See page 355).
2. Polyneuropathy (familial or acquired).
3. Distal myopathy of Gower.
4. Sometimes in myotonic dystrophy.

Causes of wasting of muscles :

- | | |
|---|-------------------------------|
| 1. LMN lesion due to any cause. | 6. Thyrotoxicosis. |
| 2. UMN lesion — There is disuse atrophy only. | 7. Malnutrition. |
| 3. Muscular dystrophy. | 8. Disseminated tuberculosis. |
| 4. Disseminated malignancy. | 9. AIDS (slim disease). |
| 5. Diabetes mellitus. | 10. Anorexia nervosa. |

* Causes of unilateral wasting of leg : old poliomyelitis, diabetic amyotrophy, disuse due to severe arthritis, prolapsed intervertebral disc, cerebral palsy.

Power testing of different muscles involved in DMD :

1. Calf muscles (gastrocnemius and soleus) — Patient lies supine with legs extended. Ask the patient to plantiflex the foot against resistance (keep the ankle fixed by holding it firmly).
2. Quadriceps — Patient lies supine; first bend the knee and then ask the patient to straighten the leg when resistance is applied over the shin.
3. Glutei — Patient lies supine; raise the patient's foot off the bed and ask him to push it down against your resistance. Knee must be kept extended. Gluteus maximus helps hip extension.
4. Deltoid—First abduct the patient's arm to 30° and tell him to keep his arm in that position. Now ask him to abduct his arm upto 90° against your resistance (supraspinatus abducts the first 30°).
5. Infraspinatus — The examiner stands behind the patient. The patient is asked to tuck his elbow into his side with the forearm placed at right angle. He is now asked to rotate the limb outwards against your resistance (elbow will be placed against the side throughout the test).

6. Latissimus dorsi — Palpate the posterior axillary folds from behind when the patient coughs. Compare the contraction on both sides; or ask the patient to clasp his hands behind his back and you offer resistance to the downward and backward movement (stand behind the patient).
7. Pectoralis major — Standing in front of the patient, ask him to clap his hands together and you attempt to keep them apart. The contraction of pectoralis can be seen.

[N.B. : Hamstrings muscles—The patient lies supine with knee flexed at 90°. Ask the patient to pull the heel towards buttock while you resist it by holding the leg at ankle.]

* **While testing the 'power' of muscles :**

Before testing, always try to demonstrate the movement rather than explaining it verbally.

- (i) Expose the muscle fully.
- (ii) Ask the patient to contract the muscle against your resistance,
- (iii) See the muscles contracting,
- (iv) Feel the strength of contraction, and
- (v) Compare with your own strength or what you judge to be normal.

** there are two methods of testing muscle power ;

- (i) **Isometric testing**— The patient is asked to contract the muscle powerfully and to maintain the contracted position while the examiner tries to keep it in original position. In isometric testing, there is no shortening of muscle.
- (ii) **Isotonic testing**— The patient is asked to contract the muscle and the examiner opposes the movement at the initial part of contraction.

*** **Isometric method is more sensitive and detects minor degree of weakness** though isotonic testing is commonly practised method in neurology.

D/D you **will consider in this case :**

1. Different causes of proximal muscle weakness.
2. Peripheral neuropathy - Sensory changes, distal involvement.
3. Motor neurone disease - Adults with asymmetrical wasting and fasciculation.
4. Post-poliomyelitis - Jerks lost with asymmetrical atrophy (characteristics of poliomyelitis—post-febrile, asymmetrical, non-progressive, without any sensory loss and with LMN signs).

How do you like to investigate myopathy ?

1. Enzyme study - Serum aldolase and CK are raised. Normal value of aldolase is 1.5-8.1 units / litre and CK is 55-170 units / litre. Enzymes are often increased by 100-200 times than normal.
2. Electromyography (EMG) shows myopathic pattern.
3. Muscle biopsy - 'Confirms' the diagnosis of myopathy. It shows islands of muscular degeneration as well as regeneration in the ocean of fibrofatty tissue, without any cellular infiltration. There is diminished dystrophin level in muscle tissue, determined by immunochemical staining.
4. CSF study and nerve conduction velocity (NCV) are normal.
5. ECG change — Tachycardia, tall R waves in right precordial leads, deep Q waves in left precordial leads, bundle branch block due to cardiac involvement (cardiomyopathy).

Do you know its management ?

No specific management of muscular dystrophy is available. Active exercise, passive physiotherapy, orthopedic measures are often helpful. Usually death occurs in second decade of life due to pneumonia, respiratory failure, cardiomyopathy or cardiac failure.

Identification points :

1. Male child.
2. Waddling gait and positive Gowers' sign (ask the patient for demonstration).
3. Pseudohypertrophy (principally the calf muscles) plus atrophy of muscles.
4. Proximal muscles weakness (ask the patient to get up from sitting position).

* **Becker type** of dystrophy (X-linked recessive) usually starts between 5-25 years. It is the late onset, 'milder' form of myopathy with slow progression as well as less cardiac involvement. The patients are chair-bound 25 years after the onset and may even survive upto 5th or 6th decade.

** DMD is caused by mutation of the gene responsible for production of the protein dystrophin.

Case 97

JERKS AND CLONUS

What is jerk, deep reflex, stretch reflex or tendon reflex ?

When a tendon is lightly stretched and struck by a single sharp blow with a soft rubber hammer, there is brief contraction of that muscle, which is known as tendon reflex. The tendon reflex is a 'monosynaptic' stretch reflex (note : superficial reflexes are polysynaptic nociceptive reflex). By elicitation of a jerk, the integrity of the afferent and the efferent pathways along with excitability of the anterior horn cells in the spinal segment of the stretched muscle are tested.

Prerequisites before elicitation of a jerk :

1. Use a hammer with firm and flexible shaft.
2. Stand on the right side of the patient (even for elicitation of jerks for the left side).
3. Explain the procedure to the patient. Reassure that it will not hurt him.
4. The patient lies supine in the bed in a comfortable and relaxed position.
5. Tap the tendon, not the muscle belly (specially in triceps jerk).
6. Expose the muscle (being tested) fully—a must for all the reflexes.
7. Observe both the contraction of the muscle and the movement of the limb.
8. Always compare with the other side and observe for the 'symmetry'.
9. Use the hammer by 'swinging movement' of the wrist joint.
10. Sudden, single and sharp blow with sufficient threshold are applied over the tendon.
11. Genitalia should be properly covered in eliciting jerks of lower limbs.
12. Always use same type of hammer in same manner (i.e., maintain the uniformity). Repeat the procedure, if necessary).

* While eliciting different tendon reflexes (see below), the position of the limb is maintained in such a way that the tendon to be tapped is slightly stretched.

Jerks commonly examined in clinical neurology :

Root values and peripheral nerves concerned in different jerks are given below :

1. Ankle jerk (S_{12})—medial popliteal nerve.
2. Knee jerk (L_{234})—femoral nerve.
3. Biceps jerk (C_{56})—musculocutaneous nerve.
4. Triceps jerk (C_{67})—radial nerve.
5. Supinator jerk (C_{56})—radial nerve.
6. Jaw jerk—trigeminal nerve.

Ankle jerk :

1. Conventional method—The patient lies supine with the lower limb (to be examined) flexed at the knee, foot slightly everted (the foot may rest on the opposite leg). Now dorsiflex the foot with the left hand and apply a sharp tap on tendo-Achilles. A quick contraction of calf muscles with plantiflexion of foot occur.
2. Special method The patient takes kneel-down position on a chair with both feet hanging out of the chair. Sharp tap is applied over tendo-Achilles on both sides in turn (do not passively dorsiflex the foot). Calf muscles contract and plantiflexion of foot results. It is done specially in myxoedema to observe the delayed relaxation time (myotonic reflex). Careful observation of delayed relaxation time in ankle jerk (due to 1 muscle metabolism) often clinches the diagnosis of myxoedema at the bedside. Myotonic reflex is classically seen in ankle jerk.

Knee jerk :

1. Conventional method—The patient lies supine with both the knee joints flexed at about 90°. Now place your forearm, dorsum of the wrist or palm under the knee (to be tested) or both the knees, and ask the patient to rest the knee completely on your hand. Be careful so that the legs should not be in contact with each other in bed. Feel the quadriceps tendon and apply a sharp tap there. The quadriceps muscle contracts with extension of the knee.
2. Special method—The patient sits on a chair with the legs hanging free side by side. After tapping the patellar tendon, look for the pendulous movement of the legs. This is classically seen in cerebellar lesion and sometimes in chorea (indicates hypotonia). Hung-up reflex (the knee may be held in extension for few seconds before relaxing) may be seen by this special method (read the section on 'Cerebellar disorder' too).

N.B. : The 'patella' hammer derives its name from knee jerk (because the patellar tendon is tapped) as it was the first tendon reflex which was regularly practised for neurological examination.

Biceps jerk :

The elbow remains semiflexed at right angle with a semipronated position of the forearm. The limb may rest on patient's abdomen or upon your left hand. Place your left thumb or index finger (transmitter for blow from hammer) on the biceps tendon in the antecubital fossa and tap suddenly over your finger. The biceps muscle contracts with flexion of the elbow.

Triceps jerk :

Place the patient's hand over his chest with the elbow flexed. Support the hand at the wrist by your left hand so that the upper limb does not fall on the bed. Suddenly tap above the olecranon process (strike the triceps tendon, not the belly of the muscle; tap 2" above the elbow) and observe the contraction of triceps with slight extension of the elbow. Muscle contraction after direct hammering is not a stretch reflex.

Supinator jerk :

Forearm is semipronated and the elbow is flexed. Hold the patient's hand at wrist and a sharp tap is applied on the styloid process which results in contraction of brachioradialis, supination of the forearm, and flexion of the elbow with minimal flexion of the fingers. As this muscle was previously called supinator longus, the reflex is known as supinator jerk (also referred as radial jerk).

Inversion of supinator jerk :

When there is a lesion in the spinal cord at C₅₋₆ segment (commonly due to cervical spondylosis resulting in myeloradiculopathy), there is hyperexcitability of the anterior horn cells below this level. So, **during elicitation of supinator jerk**, there is no flexion at elbow but only brisk flexion of the fingers (as C₇ take upperhand) occur; this is inversion of supinator jerk.

Similarly, in INVERSION OF BICEPS JERK (lesion at C₅₋₆ segment), there is no contraction of biceps during elicitation of biceps jerk but one can see the contraction of triceps (as C₆₋₇ take upperhand). Inversion of a jerk localises the level of lesion in the spinal cord. Usually, inversion of supinator and biceps jerks are seen in a single patient, and is most commonly found in cervical spondylosis.

Jaw jerk :

Prerequisites :

1. Preferably patient should be seated with mouth partially open.
2. Tongue remains inside the mouth.
3. Left thumb or forefinger is placed over the middle of the patient's chin.
4. Strike the hammer lightly on the thumb in a downward direction.

The normal response is slight and consists of sudden closure of the mouth due to contractions of masseter and temporalis. This reflex is just present or absent in health. The jaw jerk is increased in bilateral UMN lesions above the Vth cranial nerve nucleus e.g., pseudobulbar palsy or multiple sclerosis. Both the afferent and the efferent fibres pass through the trigeminal nerve, and the centre for this reflex lies in the pons. Interpretation regarding mere presence or briskness is often very difficult.

What is finger-flexion jerk ?

Its presence indicates hyperreflexia, and exaggeration or asymmetry suggests UMN lesion on the abnormal 'side'. The tips of your middle and index fingers are placed across the palmar surfaces of the proximal phalanges of the patient's relaxed fingers. A sudden tap by a hammer on your fingers produces flexion of patient's fingers, but a very brisk contraction indicates hyperreflexia. The root value of this reflex is C₇₋₈ and T_r and the peripheral nerves involved are median and ulnar nerve.

Hoffmann's sign :

Flex the DIP joint of patient's middle finger and now flick it down suddenly. The response is brisk flexion and adduction of the thumb as well as flexion of the tip of other fingers. It is also a manifestation of hyperreflexia. This reflex may not be present in all patients with pyramidal tract lesion and moreover, it may be present in a nervous individual without any organic lesion. If the reflex is seen on one side (unilateral) only, it may have some value as a sign of pyramidal tract lesion. The root value of the reflex is C₈, T_r

Wartenberg's sign :

Interlock the flexed fingers of the patient with your flexed fingers and try to pull apart. In health, the thumb remains abducted and extended but in pyramidal tract lesion, the thumb flexes and adducts.

Interpretation of tendon reflexes :

- (A) Present — In health.
- (B) Lost or diminished —
1. LMN lesion (poliomyelitis, syringomyelia, peripheral neuropathy, tabes dorsalis, peroneal muscular atrophy etc.)
 2. UMN lesion in shock stage.
- * Isolated loss of a reflex usually suggests radiculopathy. Symmetrical loss of reflex may be found in peripheral neuropathy. Reflexes may be absent if the patient is 'unable to relax'.
- (C) Exaggerated —
1. Anxiety neurosis, nervousness, hysteria.
 2. Thyrotoxicosis.
 3. Tetany.
 4. Tetanus.
- (D) Brisk —
1. UMN lesion.
 2. Sometimes in hypoglycaemic coma.
- (E) Pendular —
1. Cerebellar lesion.
 2. Chorea.
- (F) Delayed relaxation (ankle jerk) — Myxoedema.
- (G) Hung-up reflex (knee jerk) — Chorea, myxoedema, hypothermia.
- (H) Inversion of supinator jerk — Level of lesion in C₅₋₆ spinal segments

** Hyperreflexia means exaggerated or brisk response. For a student it is very difficult to differentiate between the two. In a broader sense, exaggerated reflex means the 'amplitude' of the limb movement is more and brisk reflex means, the reflex is very 'prompt' in its response. We may conclude that hyperreflexia is only of pathological significance when it is asymmetrical (comparing with the other side) or if associated with other signs of UMN lesion (spasticity, Babinski's sign etc.)

*** A reflex seems lost or diminished in defect of technique, relaxation, or observation.

**** Sometimes, lesion in the spinal cord may give rise to 'crossed reflex induction', e.g., while eliciting a knee jerk on right side, it may produce a reflex response on the left knee jerk.

What is Jendrassik's manoeuvre ?

It is also known as 'reinforcement'. When the deep reflexes or jerks are not elicited by routine manoeuvre, reinforcement is applied for augmentation of response. It is done by distraction of patients' attention. Ask the patient,

1. To make a fist with the contralateral hand, or
2. To interlock the fingers of two hands together and then pull them as hard as possible, or
3. To clench the teeth, or
4. To push the knees hard together.

It is very important to remember that this phenomenon lasts for less than a second. So the patient is asked to do the manoeuvre almost synchronously with the tapping of the tendon (the procedure must be demonstrated to the patient before tapping the tendon). Reinforcement (to make some strong voluntary muscular effort) acts by,

- a) Increasing the excitability of the anterior horn cells, and
- b) Increasing the recruitment of gamma-fibres, i.e., increasing the sensitivity of the muscle spindle primary sensory endings to stretch (increased gamma fusimotor drive).

* **Always perform the Jendrassik's manoeuvre before declaring a tendon reflex 'absent'.**

** For upper limb reflexes—1, 3, 4; for lower limb reflexes—2, 3.

Causes of loss of knee jerk :

It signifies the lesion at the level of L₂₋₃₋₄ spinal segment. The causes are :

1. Non-cooperative patient or faulty technique.
2. Acute anterior poliomyelitis.
3. Peripheral neuropathy.
4. Myelopathy in shock stage.
5. Progressive muscular atrophy (MND).
6. Subacute combined degeneration.
7. Friedreich's ataxia.
8. Tabes dorsalis.
9. Sometimes in diseases of myoneural junction (e.g., myasthenic crisis).
10. Muscular dystrophy in late stages.
11. Ankylosis of the knee joint.

* In case of loss of deep reflex restricted to one limb only, one should not forget acute anterior poliomyelitis, entrapment neuropathy, leprosy and diabetic mononeuropathy.



Chronic tophaceous gout with tophi present over olecranon process, tibial tuberosity, shins, feet, tendo-Achilles and dorsal aspect of fingers



Ladder-pattern **visible peristalsis** in acute small intestinal obstruction



Spider naevi at upper part of back in cirrhosis of liver



Rheumatic nodules – small nodules over olecranon process of a young girl suffering from acute rheumatic fever



Discoid lupus erythematosus (DLE)



Ascites with umbilical hernia. Abdominal paracentesis should be done to avoid strangulation of hernia



Hands in scleroderma showing sclerodactyly (tight skin over fingers), pulp atrophy (acrosclerosis in right index finger), digital infarction, nail changes, salt-pepper skin and H/O Raynaud's phenomenon



Carcinoma of left breast with metastatic cervical lymphadenopathy and **lymphoedema** of left upper extremity



Alopecia areata in scalp



Vitiligo

What are the different responses ?

Usually there are 5 types of responses :

1. Flexor.
2. Extensor (up-going toe).
3. Equivocal.
4. No response.
5. Withdrawal response.

Briefly describe the different responses :

(A) **Flexor**—Normal response; already described.

(B) **Extensor** (extensor plantar response is also known as "Babinski's sign"). The responses are :

1. Dorsiflexion (extension) of the great toe (movement occurs at the metatarsophalangeal joint) which usually precedes all other movements mentioned below.
2. Fanning (spreading out) and extension of outer four toes.
3. Dorsiflexion of the ankle, and flexion of the knee and the hip.
4. Contraction of tensor fascia lata.

* In 1896, Babinski first described this sign. Josef Francois Felix Babinski (1857–1932) was a French physician cum neurologist. This sign is regarded as THE SIGN in clinical neurology.

(C) **Equivocal**—i.e., incomplete response or variable response. When the full components of the extensor response is not manifested e.g., (i) only fanning of outer four toes is seen without any movement of the great toe, or (ii) the hemiplegic side does not show any response and the healthy side shows flexor response (sometimes seen in early cases of CVA), or (iii) asymmetry of 'flexor' response in both sides.

Remember, today's equivocal response may be tomorrow's extensor response.

(D) **No response**—After striking the sole of the foot, there is no movement of any of the toes.

(E) **Withdrawal response**—Often withdrawal response is seen in anxious individuals or in normal persons with hyperaesthetic / sensitive sole. It is seen that the initial normal flexor response is quickly followed by mass extension of toes with withdrawal of the entire leg.

** The afferent nerve of this reflex (plantar response) is tibial nerve; the efferent nerve is tibial nerve for flexor response and peroneal nerve for extensor response.

*** Babinski's sign is the most sensitive, delicate and reliable sign of corticospinal tract lesion.

**** In hemiplegia or paraplegia (i.e., UMN lesion), the plantar response remains extensor life-long.

***** 'Crossed' extensor response—on unilateral stimulation of the sole in a case of bilateral cerebral or spinal cord lesion, bilateral extensor response can be evoked (e.g., in severe spastic paraplegia).

Significance of presence of Babinski's sign :

It signifies pyramidal tract lesion with or without anatomical discontinuity. The causes are :

1. **Pyramidal tract lesion** (i.e., UMN lesion above the S₁ level of the spinal cord).
2. Children below 1 year of age (due to lack of myelination of pyramidal tract).
3. Deep coma due to any cause.
4. Deep sleep.
5. Deep anaesthesia; neuroleptics or narcotic overdose; hypnosis; drugs (e.g., barbiturates).
6. Metabolic encephalopathy.
7. Often in hypoglycaemia (even in the absence of coma).
8. Immediately after the attack of generalised convulsions or epilepsy (post-ictal phase).
9. Electroconvulsive therapy.
10. Physical exhaustion (e.g., marathon runner).
11. Apnoea phase of Cheyne-Stokes respiration.

N.B. : Except 1, all are the causes of lesion in pyramidal tract without anatomical discontinuity, and thereby produce transient plantar extensor response. No. 8, 9 and 10 are due to exhaustion of corticospinal tract.

'Reinforcement test' of plantar response, if any :

1. Rotating the patient's head to opposite side.
2. Knee must be extended during the test.
3. Apply warmth to the cold skin of the sole.

Causes of delayed relaxation of ankle jerk :

1. Myxoedema (the contraction phase is delayed too) — Commonest cause
2. Gross pedal oedema
3. Hypothermia
4. Obesity
5. Parkinsonism.

Grading of reflexes :

0	=	Absent		Exaggerated or hyperactive +++
1	=	Present (as normal ankle jerk)		Normal ++
2	=	Brisk (as normal knee jerk)	Or	Diminished +
3	=	Very brisk		Absent -
4	=	Clonus		Only with reinforcement ±

Muscles acting of different joints :**(A) Ankle joint :**

- a) Dorsiflexion—Tibialis anterior
- b) Plantiflexion—Gastrocnemius and soleus (calf muscles)
- c) Inversion Tibialis anterior and tibialis posterior
- d) Eversion—Peroneii muscles

(B) Knee joint :

- a) Extension—Quadriceps femoris (i.e., three vasti + rectus femoris)
- b) Flexion Hamstrings muscles (biceps femoris, semimembranosus and semitendinosus)

(C) Hip joint :

- a) Extension—Gluteus maximus mainly
- b) Flexion—Ileopsoas
- c) Adduction—Adductor muscles
- d) Abduction—Gluteus medius and minimus

What is clonus ?

It is the rhythmical contraction of a muscle or group of muscle in response to sudden passive and sustained stretching. Clonus is always associated with brisk tendon reflex, spasticity and Babinski's sign. It is a very reliable sign of pyramidal tract lesion. There are two types of clonus •

1. Sustained clonus (true clonus).
2. Unsustained clonus (pseudoclonus).

Sites for clonus in clinical neurology :

1. Ankle clonus.
2. Patellar clonus (or knee clonus).
3. Jaw clonus (elicit the jaw jerk to see a series of closure and opening of the mouth).
4. Wrist clonus (elicited by sudden passive hyperextension of the wrist which will evoke clonic movement of palm).

N.B. : No. 3 and 4 are not routinely practised in clinical neurology.

Ankle clonus :

Patient lies supine. Support the **flexed knee** (both knee and ankle resting in 90° flexion) with your left palm by placing it in popliteal fossa in such a way that heel rests gently on the bed. Now suddenly dorsiflex the foot by grasping the forepart of it with your right hand. A series of contraction and relaxation of calf muscles are seen if a steady pressure is maintained on the foot by the right hand.

Patellar clonus :

Patient lies supine with **knee extended**. Patella is pulled upwards by the index finger and the thumb with a fold of skin behind the palm. Now a sharp and sudden displacement of the patella downwards with sustaining the pressure will produce series of quadriceps contraction.

N.B. : Ankle and patellar clonus are produced as a result of loss of upper motor control (i.e., UMN lesion) over $S_{1,2}$ and $L_{2,3,4}$ spinal segments respectively.

Pseudoclonus :

1. The clonus is not sustained inspite of a steady pressure.
2. Irregular in rate (usually less than 6 at a time) and rhythm.
3. It is commonly seen in anxiety, in tense persons, often in hysteria and thyrotoxicosis.

4. Plantar response is always flexor.

* Many a time, the boundary between true and pseudoclonus is not very clear.

Conclusion :

If you are in doubt whether the clonus is true or pseudo, immediately do the following :

1. Plantar response will be extensor in true clonus and flexor in pseudoclonus.
2. Examine the pulse (high volume in thyrotoxicosis, tachycardia in anxiety) and look for (metabolic) tremor (anxiety, thyrotoxicosis) — if present, indicates pseudoclonus.

* **Never forget to examine a patient for clonus if there is presence of brisk tendon reflex.**

Case 98

Plantar Response (Reflex)

Method of elicitation :

One should learn how to elicit this superficial reflex properly. At first, educate the patient about the procedure. The patient lies supine with extended legs and relaxation of the muscles of the lower limbs. Hold one leg firmly above the ankle joint with your left hand (to prevent withdrawal response) in such a way so that toes of the foot look towards the roof, and the outer border of the sole is scratched gently but firmly (neither too speedy nor too slow) by the tip of a key (preferable stick of the hammer or blunt needle. Starting from the heel, go along the lateral border towards the little toe and then turn medially across the metatarsal pad up to the head of 2nd metatarsus (**never touch the ball of the great toe and the flexor creases of the toes**) in a semicircular fashion. The stimulus should not produce injury but it should be of noxious character (a combination of pressure pain and touch) since this is a 'nociceptive reflex'. Stop stimulating the sole as soon as you get the first movement of the great toe. Now do the test on the other side. Plantar response is a spinal reflex arc modified by the pyramidal tract.

It is the most important and most frequently misinterpreted reflex in the body.

What is the normal plantar response ?

The normal response of this age-old sign in neurology is 'flexor' in type and is manifested by :

1. Flexion of all the five toes.
2. Dorsiflexion and inversion of the foot (with a stronger stimulus).
3. Contraction of adductors of the thigh, sartorius and tensor fascia lata (minimal response).

Precautions taken before eliciting plantar response :

1. Assess the thickness of the sole—Thick sole may be responsible for 'no response'.
2. Look for any deformity of great toe—Move the great toe, and confirm that it is moving freely and is not rigid (e.g., hallux rigidus, rheumatoid arthritis).
3. Knee must be extended.
4. Sole should be made warm (e.g., by rubbing with your palm, specially in winter season).
5. Leg should lie straight (don't allow rotation of thigh).
6. One may assess the power of extensor hallucis longus before doing the test (optional).
7. Ask the patient whether sensation in sole is intact or not (by touching the sole with your palm).

What is the root value of this reflex ?

S..

Why the lateral aspect of the sole is stimulated ?

1. Probably the receptors for extensor plantar response are present there in abundance and thus, even in minimal UMN lesion, extensor plantar response is obtained by stimulating the lateral aspect of the sole.
2. If the medial aspect of the sole is stimulated, it may elicit flexion (even in the presence of UMN lesion) of all the toes as part of the grasp reflex, itself a different phenomenon.

No response' or loss of plantar response :

- | | |
|----------------------------|---|
| 1 Faulty technique. | 5. Peripheral neuropathy. |
| 2 Thick sole | 6- Myelopathy involving S ₁ segment. |
| 3 Coldness of the feet. | 7. Paralysis of dorsiflexors of the foot. |
| 4. Stage of neural shock'. | 8- Hallux valgus or hallux rigidus. |

What are the 'plantar equivalence' ?

In case of widespread and severe UMN lesion, the reflexogenic zone may be greatly widened and an "extensor" plantar response may be evoked by different techniques. They are :

1. **LAU's sign**—Heav pressure is applied by the thumb and Index finger from above downwards to anterior surface of tibia (to its medial side). The extensor response usually

occurs towards the end of the stimulation.

2. **Gordon's reflex**—Squeezing or applying deep pressure to calf muscles produce extensor plantar response.

3. **Shefer's sign**—Extensor plantar response is evoked after applying deep pressure to tendo-

Achilles.

4. **Chaddock's sign**—Extensor response is seen after striking the skin around the lateral malleolus in a circular fashion.

5. **Bing sign**—Pricking the dorsum of foot or the great toe by a pin produces extensor response.

6 **Moniz sign**—Extensor response is seen after forceful passive plantar flexion of ankle

7. **Gonda sign**—Extensor response is elicited after forceful stretching or snapping of distal phalanx of either of the 2nd or 4th toe.

8 **Brissaud's reflex**—Contraction of tensor fascia lata as a part of extensor response (palpate the upper and lateral part of thigh for appreciation). This is helpful in patients *with amputated or absent great toe, or complete paralysis of extensors of toes.*

* 123 and 4 are commonly practised methods in clinical medicine.

- These reflexes are useful in non-cooperative patients or when the soles are extremely sensitive or the soles are wounded/injured.

What is Rossolimo's sign ?

Method : Either tap the ball of the foot (by percussing the plantar surface of the ball of the great toe with a hammer) or flick the distal phalanges of the toes into extension and then allow them to back into their normal position.

Response : Pyramidal tract lesion, manifested by plantiflexion of all the five toes (only sign with UMN lesion, manifested by plantiflexion of great toe). It is the homologue of Hoffmann's sign of upper limb.

Plantar response is extensor with loss of ankle jerk :

1. Subacute combined degeneration (SCD) of the spinal cord.
2. Taboparesis (GP1 plus tabes dorsalis).
3. Friedreich's ataxia.
4. Cauda-cona lesion (cauda equina lesion with lesion in conus medullaris).
5. Combined cervical and lumbar spondylosis.

Plantar response is flexor with brisk ankle jerk :

1. Sometimes in anxiety and thyrotoxicosis (exaggerated deep reflex).
2. Hepatic pre-coma.

Why the ball of the great toe is not touched while eliciting plantar response ?

During the elicitation of plantar response, one should stop scratching the sole at the head of 2nd metatarsus. If the ball of the great toe is scratched, there may be dorsiflexion of the great toe (by direct stimulation) in the absence of any UMN lesion. It is why the root value of the plantar response is only the 1st sacral segment of the spinal cord.

N.B. : *The first movement of the great toe is important.* Extension of great toe after a brief initial flexion is not extensor response. Remember, there is nothing called negative or positive Babinski's sign.

Pseudo-Babinski's sign may be seen in plantar hyperaesthesia or chorea.

Case 99

TROPHIC CHANGES (NEUROGENIC)

What are the trophic changes ?

1. Trophic changes in the skin and appendages,
2. Trophic ulcers, and
3. Charcot joint (neuropathic joint).

Changes in the skin and appendages :

- | | |
|------------------------------------|-------------------------------------|
| 1. Dry, rough and cold skin. | 4. Hypohydrosis (lack of sweating). |
| 2. Pigmentation. | 5. Hypotrichosis (fall of hairs). |
| 3. Local cyanosis or local oedema. | 6. Brittle nails. |

Sites examined for trophic changes :

Denudation of skin with ulcer (bed sore) formation over the bony prominences is seen :

- | | |
|------------------------------|--|
| 1. On the lateral malleolus. | 3. Shoulder girdles. |
| 2. Back of the heel of foot. | 4. Over the sacrum (classical bed sore). |

* Bed sore may form in a patient with prolonged recumbency, specially without change of posture.

What is trophic ulcer :

These perforating ulcers are usually present **over the heel or ball of the great toe** During walking, heel is touched first in the ground and thus, it is a common site for pressure sore formation. The patients with foot drop touches the ground with the ball of the toes (leprosy, diabetes mellitus) and thus, trophic ulcer forms there. In syringomyelia and tabes dorsalis, the ulcer is usually situated in the heel (as no foot drop is there). Trophic ulcer is usually painless.

* Tropical ulcer' (totally different from trophic ulcer) is a chronic form of callous ulcer with its edge raised and undermined. It often refuses to heal.

Common causes for trophic ulcer in feet :

Same as the causes of Charcot joint (see page 484).

* Peripheral nerve injury (in legs) and spina bifida may give rise to trophic ulcer in feet.

Aetiopathogenesis of trophic changes :

1. Loss of muscular action upon the circulation,
2. Vasomotor paralysis due to destruction of the vasoconstrictor (sympathetic) fibres, and
3. Partly due to disuse.

* **Search for trophic changes in hemiplegia, paraplegia and peripheral neuropathy.**

** **Autonomic nervous system (ANS)** may be involved in hypothalamic disorders, diabetes mellitus or spinal lesions. The points noted during examination of ANS are :

Sleep disorder (insomnia or somnolence), temperature regulation, genital dysfunction (impotence or amenorrhoea), sweating (diminished or excessive), postural or orthostatic hypotension, trophic changes (e.g., ulcer), Horner's syndrome, bladder and bowel dysfunction (urinary incontinence, diarrhoea), pupillary areflexia.

Points to note in examination of cranium and spine in neurology :

(A) Cranium (inspection, palpation, percussion and auscultation) —

1. Microcephaly (cerebral palsy), macrocephaly (hydrocephalus. Paget's disease) or turricephaly (tower-like peaked appearance due to synostosis of sutures), dolichocephaly (long antero-posterior diameter), brachycephaly (short antero-posterior diameter) or oxycephaly (cone-shaped and pointed head).
2. Haematoma, elevation, fracture or depression, scar mark, secondaries in skull, mass in myeloma.
 3. Fontanelle (tense fontanelle is seen in hydrocephalus and raised intracranial tension).
4. Sutures (springing of the sutures is seen in hydrocephalus).
5. Cracked-pot sound in hydrocephalus on percussion.
6. Bruit (in angiomatous malformation or stenosis)—Carotid artery bruit in neck at carotid bifurcation, vertebral or subclavian artery bruit at supraclavicular fossa, and bruit in cerebral arteriovenous malformation may be heard over cranium or closed eye.

(B) Spine —

1. Any tenderness.
2. Gibbus (e.g., caries spine, metastasis, trauma).
3. Kyphoscoliosis.
4. Spina bifida occulta (by inspecting the tuft of hair, dimpling of skin, fibrofatty tumour or dilated vessels over the sacrum).
5. Short neck (cranio-vertebral anomaly).
6. Bruit heard over spine in angiomatous malformations.

* **One must examine the spine in all neurological cases, specially in paraplegia.**

Case 100

CHARCOT JOINT

What is Charcot joint ?

This is a chronic, progressive degenerative arthropathy which is also known as **neuropathic joint**. The characteristics are :

1. Huge swelling of knee, hip, ankle or shoulder joint (**most common site is knee joint**).
2. *Painless (most characteristic).*
3. Disorganised, deformed and destroyed joint.
4. Increased mobility (rarely, genu recurvatum may be produced) of the joint and thus, easy dislocation is possible—unstable or subluxed joint.
5. Loose bodies may be palpated in the joint cavity and crepitus may be felt with new bone formation (osteophytes).

What are the common causes of neuropathic joint ?

1. Leprosy (commonly affects the upper extremity).
2. Diabetes mellitus (tarsal and metatarsal joints mostly affected).
3. Syringomyelia (shoulder, elbow and wrist joints mostly involved).
4. Tabes dorsalis (knee, hip and ankle joints commonly affected).
5. Sometimes found in hereditary sensory neuropathy or after repeated intra-articular injection of steroid.

Most important neurological sign tested in a patient of Charcot joint :

Posterior column sensations, i.e., joint, position, muscle, vibration and pressure senses are lost here. Charcot joint is the complication of chronic loss of proprioception.

Pathophysiology of development of Charcot joint :

There are two theories—

1. Loss of proprioception leads to recurrent trauma damage goes unnoticed by the neuropathic patient -> ultimately leads to progressive destruction, degeneration and disorganisation of the joint.
2. Hyperaemia results from neurally mediated vascular reflex -> leads to increased osteoclastic bone resorption.

Basic principles of testing sensory functions :

Sensory evaluation is the most unreliable part of neurological examination as it is subjective and is difficult to quantify. Basic principles of testing are :

1. Explain the patient clearly what is going to be tested; patient's cooperation and alertness are essential. Try to gain confidence by proper understanding.
2. *First test with eyes open and then with eyes closed.*
3. Always compare the sensory function with the opposite side for symmetry.
4. First apply the sensory stimulus to the area of altered sensation and delineate its borders by testing from abnormal to normal area. Ask the patient to say 'yes' if he feels anything.
5. A hairy area may be shaved before testing.

6. Test the spinal segments or dermatomes sequentially (e.g.. In the lower limb, first examine the L₁ segment and then proceed downwards for L₂₋₅ segments and again upwards for S segments present in different places in the said limb).

* Cervical, (CJ has no cutaneous supply, which only supplies meninges.

Superficial sensations (spinothalamic functions) tested are—

- (A) TOUCH—fine touch is tested with a small piece of cotton wool which is twisted into fine hair while crude touch may be tested by the wider side of a cotton wool or the tip of index finger. Fine touch sensation may be evaluated over toes, metatarsal heads, heels, and dorsum of feet with a 10 g monofilament (Semmes Weinstein monofilament), specially in a patient of diabetes.
- (B) PAIN—usually tested by a pin (avoid heavy pressure); select the pre-sternal area for baseline sharpness before testing a limb; ask whether the quality of sensation becomes sharper or painful (hyperaesthesia). or feels blunter (hypoesthesia).
- (C) TEMPERATURE—actually it is tested by two test tubes containing cold (5°-10°C) and hot (37°-45°C) water touched on face, forearms, hands, trunk and legs in a random sequence; a rough assessment is done by the metal of tuning fork or stethoscope (cold), or by rubbing the palms (hot) as a clinical bedside routine. A normal person can appreciate even a difference of 1°C.

* Deep pain sensation is tested by pressing the calf muscles, tendo-Achilles or testes.

How to test for deep and cortical sensations ?

Deep sensations (posterior column functions) tested are—

- (A) VIBRATION SENSE—it is the test of repeated touch sensation in quick succession. First keep the vibrating tuning fork (128 cycles/sec) over the sternum so that the patient can identify the sense of vibration (not the touch sensation). On a particular site, vibration sense should be perceived minimally for 10 seconds. Instruct the patient to close his eyes. For the lower limb, hold the vibrating fork sequentially over the great toe, dorsum of foot, medial malleolus, tibial shaft (shin) and tuberosity, and anterior superior iliac spine. In the upper limb, the fork is kept sequentially over DIP and PIP joints of forefinger, dorsum of hand, wrist joint, olecranon process and over the shoulder. In the trunk, it is tested by placing the fork over ribs, sternum, clavicles and vertebral spines. Always compare with the other side. From time to time, judge the patient by stopping vibrating fork with your hands. The rule goes like this—if the distal vibration sensation persists, it is useless to examine the proximal parts but in case of loss of distal sensation, always move proximally in turn. Vibration sense is commonly lost in posterior column lesion (tabes dorsalis), peripheral neuropathy (specially, diabetes mellitus) and old age (physiological).
- (B) MUSCLE SENSE—compress or squeeze big muscle bellies (calf, triceps or biceps) and note whether the patient complains of pain (Abadie's sign) or not. This is the test of PRESSURE SENSE too.
- (C) JOINT AND POSITION SENSE—first, explain the patient about the test with movements of joints ('it is up' or 'it is down'). Now, ask the patient to close the eyes. For examination of the lower limb, grasp the proximal phalanx of great toe by your left hand, and hold the **medial and lateral borders of the distal phalanx** with the thumb and index finger of your right hand so that pressure above and below does not reveal the direction of movement. Move the distal phalanx up and down at random (15°-30°) and ask the patient whether any movement is perceived by him or not (joint sense), and to identify the direction of movement (position sense). In health, movements of < 10° can be perceived at all normal joints. In the upper limb, this test can be done in the index finger. Perform the test on the other limb too. If there is impaired joint sense in small joints, test the big joints in turn. Caution should be taken during the test so that your fingers should not rub against the patient's other toes. At least four wrong answers should be received before declaring the joint sensation impaired or lost. Position sense can also be tested by instructing the patient to keep one limb in a particular position and then ask him to keep the opposite limb in the similar position.

Cortical sensations (parietal lobe functions) tested are—

- (A) ONE POINT LOCALISATION—ask the patient to localise accurately parts of his body touched by a cotton wool (eyes remain closed).
- (B) TWO POINT DISCRIMINATION—it is tested by the use of blunt compass or divider. In health, a minimum distance of 2-3 mm on the lips, 3-5 mm on the finger tips, 2-3 cm on the dorsum of hands, or 3-4 cm on the dorsum of the foot can be recognised as two different stimuli.

- (C) STEREOGNOSIS—small objects of various size e.g., coins, match box, pencil, key (i.e., familiar objects) are placed in the patient's palm and asked to identify while the eyes remain closed. Recognition of size, shape, weight and form of a common object, and identification of it by touch and manipulation is known as **stereognosis**. Patient's failure is known as astereognosis.
- (D) GRAPHAESTHESIA—draw letter, number, diagram or circle with a blunt object, or with your index finger over the patient's back, thigh or forearm (eyes remain closed). A normal person can easily identify the letter or number written on the skin surface (graphaesthesia).
- (E) SENSORY EXTINCTION (SENSORY INATTENTION, SENSORY NEGLECT, PERCEPTUAL RIVALRY)—homologous points on two sides of the body are pricked with a pin separately (eyes remain closed). If the patient can identify the pin prick in both the situations, the previous points are now pricked 'simultaneously'. In unilateral parietal lobe lesion, the sensation on the opposite side is not perceived by the patient.

* Cortical sensations are carried to contralateral parietal lobe through peripheral nerves via posterior column. If peripheral nerves or primary sensations are at jeopardy, it is useless to test cortical sensations.

** See page 176 for different sensations carried through spinothalamic tract and posterior column.

What are the patterns of different sensory disturbances ?

Sensory modalities should be tested in search of a pattern consistent with spinal cord, spinal root or peripheral nerve abnormalities.

1. Peripheral neuropathy— Symmetrical 'glove and stocking' paraesthesia or anaesthesia, affecting the distal parts more; involves all modalities of sensations.
2. Nerve root lesion— Pain is often felt more than the loss of touch sensation; dermatomal pattern of sensory loss in the distribution of nerve roots.
3. Complete section of the spinal cord— Loss of all modalities of sensations below the level; a narrow zone of hyperaesthesia on the top of anaesthetic zone. 'Sacral sparing' may be present in few cases of high cord compression.
4. Hemisection of the spinal cord (Brown-Sequard syndrome)— Vide page 160.
5. Cauda cona lesion— 'Perianal anaesthesia', i.e., all modalities of sensations are lost involving lower sacral segments. In cauda equina lesion, there is asymmetric sensory loss in lower limbs.
6. Anterior spinal syndrome— e.g., in anterior spinal artery thrombosis there is loss of pain, touch and temperature on both the sides below a level, with preservation of proprioception.
7. Posterior spinal syndrome— It is just the opposite of the anterior spinal syndrome. There is loss of joint, position and vibration sense below a particular level with preservation of touch, pain and temperature. It is commonly seen in tabes dorsalis.
8. Central cord lesion— e.g., in syringomyelia, there is affection of spinothalamic tract due to involvement of fibres crossing the cord from both sides in the anterior commissure though it spares the posterior column. Thus, touch sensation persists but pain and temperature sensations are lost (dissociated anaesthesia). The sensory loss is in shawl-like distribution ('sleeve jacket' anaesthesia).
9. Mid-brainstem lesion— Ipsilateral sensory loss in face with contralateral loss on the body. There is loss of touch, pain and temperature sensations as a result of affection of trigeminal tract or nucleus, and lateral spinothalamic tract.
10. Thalamic lesion— Contralateral loss of all modalities of sensations (position sense is affected more) on one side of face and body. There may be history of spontaneous, disabling type of pain i.e., hyperpathia (thalamic pain).

* **Dissociated sensory loss** (a sign of spinothalamic tract involvement with sparing of posterior column), i.e., loss of pain and temperature with preservation of touch (fine) sensation is found in :

1. Anterior spinal artery thrombosis.
2. Syringomyelia, haematomyelia, hydromyelia, intramedullary tumour.
3. Posterior inferior cerebellar artery syndrome (or lateral medullary syndrome).
4. Brown-Sequard syndrome (hemisection of spinal cord).
5. Leprosy neuropathy (small fibre neuropathy).

What other signs will you search in a patient with Charcot joint ?

Always search for trophic ulcers in the heel or ball of the great toe in a patient with Charcot joint.

Radiological features of Charcot Joint :

- 1 Osteopenia (bone density is less). 3. Severe disruption and disorganisation of the joint.
2. Sharp cortical margin. 4. Inappropriate new bone formation.

Differential diagnosis of a swollen knee joint :

- | | |
|---|--------------------------------------|
| 1. Septic arthritis. | Tuberculosis of the knee joint. |
| 2. Osteoarthritis. | 8. Charcot joint. |
| 3. Huge joint effusion (e.g., traumatic). | 9. Reactive arthritis. |
| 4. Gout or pseudogout. | 10. Rheumatic fever. |
| 5. Haemophilic joint. | 11. Seronegative spondylarthritides. |
| 6. Rheumatoid arthritis. | 12. Psoriatic arthritis. |
- * Except 8, all are painful.

Management of Charcot joint :

1. Treatment of the underlying disorder.
2. Stabilisation of the joint is done by braces and splints.
3. In case of diabetes mellitus, prevent weight bearing in the foot for 2 months to prevent severe disease (trophic ulcer etc).
4. Lastly, fusion of a very unstable joint may be done.

Conclusion :

In a suspected patient of Charcot joint,

1. Test the joint for pain sensation (by moving the joint).
2. Test for other posterior column sensations.
3. Always search for trophic ulcers in foot.

Case 101**ABNORMAL GAIT****What is gait ?**

It is the posture of the patient during walking (decubitus means posture of the patient in bed).

Precautions taken before examination of gait :

The pattern of gait very often gives a clue to neurological disorder. Normal body posture and locomotion require integration of motor and sensory system, extrapyramidal system visual information, intact labyrinthine function and proprioception-the last one is the most important. One should ascertain the following facts before the examination of gait :

- 1 Examine the stance. Is the patient able to walk ? Any deviation ? Any tendency to fall
 2. Exclude the disease of joints and bone or surgical causes producing disorders in gait (say, osteoarthritis of the hip).
 3. Legs should be adequately exposed (i.e., at least upto the knee).
 - 4 Feet should be bare.
 - 5 The patient is asked to walk freely in the room, and then along a straight line (crack or grout placed in floor-mosaic); and then to turn and walk back in the original position.
 6. The patient may be asked to walk on toes or on heels. Tandem gait (heel to toe gait) should always be examined.
 - 7 Assure the patient of his safety by your close presence.
- * During walking look meticulously for posture (e.g., stooped), arm swinging, as well as involuntary movements, pain complained, fall towards one side, symmetry of steps and width of the base.

What is stance :

- 'Heel-rip r.f. of the patient on standing'. The bipedal stance is examined by asking the

ZSZ

Set to stifle toe U at a time, then to 'stand on toe,, and then on heels. The patient, may be

pulled abruptly backwards from the shoulders while noting his balance (i.e., postural reflex). Stance in different disease are :

1. Hemiplegic stance—the patient stands with extended and adducted lower limb with plantiflexed foot on the affected side, keeping the body weight on the healthy side. The affected upper limb is typically flexed, adducted and semipronated; there is flexion of the wrist, MCP and IP joints with slight adduction of the thumb.
2. Wide base stance—the patient stands with legs and feet placed wide apart to keep balance of the body weight, and is classically seen in alcoholics, sensory ataxia and peripheral neuropathy.
3. Stance with unilateral cerebellar disease—The affected-sided shoulder is held at a lower level and the body weight is thrown on the healthy side. 'Head tilt' is observed towards the affected side.
4. Stance in parkinsonism—the patient stands in a general attitude of flexion (typical 'stoop'). The head is slightly flexed on the trunk and there is general flexion of the spine. All the limbs are held in flexion in all the joints and the hands may exhibit static tremor.
5. Lordotic stance—In proximal muscles weakness (e.g., myopathy), the patient can stand erect with an exaggerated lumbar lordosis. In weakness of erector spinae muscles (e.g., spinal muscular atrophy), the patient stands with a lordotic stance.
6. Hysterical stance—The patient starts swaying which starts in hips instead of ankles but never falls on the ground. The effects are same whether the eyes are open or remain closed (false positive Romberg's sign).

Different types of gait :

1. HEMIPLEGIC or SPASTIC GAIT : This is seen in hemiplegic patients (UMN lesion) after recovery. The patient walks on a narrow base, the hemiplegic limb is held stiffly, and does not flex at the knee and hip. While the patient drags his affected foot, the foot is raised from the ground by tilting the pelvis and the leg is swung forward forming a semicircle or an arc—known as 'circumduction' of the leg. The outer side of the sole of the shoe is worn (as there is plantiflexion on the affected side). The affected arm is carried in a flexed position and does not swing naturally. The hemiplegic gait is essentially a spastic gait.
2. SCISSORS or SPASTIC PARAPLEGIC GAIT : The patient stands with crossed legs (often with a stick in one hand). Each leg is advanced slowly and stiffly with restricted motion at the knee and hip. He steps one limb in front of the other in a semicircular fashion ('scissoring' of gait). It is found in patients with spastic paraplegia and cerebral diplegia.
3. STAMPING GAIT : This is the gait of posterior column lesion or sensory ataxia, and is classically seen in tabes dorsalis. The patient stands on a broad base with careful watch on the ground. He raises his foot suddenly to abnormally high level (as he does not know where the foot is), jerks them forward and bring them on the ground with a stamp (the heel touches the ground first and one may find a trophic ulcer in the heel). These patients are severely ataxic with eyes closed. Romberg's sign is present in patients with stamping gait.
4. HIGH-STEPPING GAIT or EQUINE GAIT : This type of gait is found in patients with foot drop and is commonly seen in peripheral neuropathy, acute anterior poliomyelitis or common peroneal nerve palsy. This is just like the gait of sensory ataxia where the patient elevates his legs to a high level and bring them on the ground with a slapping noise. As there is foot drop, toes touch the ground first (trophic ulcer is found in the ball of the great toe) though a high-stepping attitude is adopted by the patient in order to avoid injury to the toes.
5. FESTINANT GAIT : It is the gait of parkinsonism (read the section on 'Parkinsonism'). It is worthwhile to remember that a patient of parkinsonism may run better than walk or walk backwards better than forward.
6. WADDLING GAIT : The walking becomes a 'waddle' i.e., the pelvis being poorly supported by each leg. Read the section on 'Myopathy'.
7. REELING, STAGGERING, DRUNKEN or ATAXIC GAIT : This is observed in cerebellar ataxia and alcoholic intoxication. The patient walks on a broad base, the unsteady feet are planted widely apart and placed irregularly. The patient sways from side to side. The steps are uncertain, some are shorter and some are longer than intended, and the patient tends to fall or deviate to the side of cerebellar lesion. The ataxia is equally severe whether the eyes are open or closed. Tandem walking is virtually impossible. Read the section on 'Cerebellar disorder'.
8. PROSTHETIC GAIT : This is the gait with an artificial limb.

9. **LIMPING GAIT** : This type of gait is adopted due to joint diseases (tuberculosis of the hip), talipes equinovarus or any painful condition in the lower extremities.
10. **HYSTERICAL or FUNCTIONAL GAIT** : The patient often walks with bent knees or trunk. This is a bizarre gait with combination of all the varieties (hemiplegic, paraplegic, ataxic etc.) and does not resemble any known pattern of organic disease. The patient drags the affected leg with an exaggerated delay, while the typical circumduction is absent; but walks normally in the absence of an observer. He/she never falls during walking. Hysterical hemiplegia patient should be examined for (to differentiate from organic hemiplegia) :
 - a) The patient lies supine and is asked to raise one leg against resistance. In a normal person, the back of the heel of the contralateral leg is pressed firmly down in the bed (examiner's hand placed under the heel), and the same is true in a patient with organic hemiplegia when he/she tries to lift the paralysed or weak leg against resistance. The hysteric will contract the good leg more strongly than as a primary' willed action (Hoover's sign).
 - b) Babinski's leg flexion test—If a patient with organic hemiplegia is asked to sit (from supine position) against resistance without using his/her arms, the paralysed leg flexes involuntarily owing to lack of stabilisation of joints while in hysteria the normal leg is flexed first.
 - c) 'Astasia-abasia'—Though the hysterical hemiplegic patient is unable to stand or walk without assistance, he/she uses his legs normally while in bed.

* 'Marche a petipa' (after the Russian ballet master)—In bilateral corticospinal lesions (diffuse cerebrovascular disease) short, stamping steps occurs with bilateral spasticity and extensor plantar response. The rapid steps resemble a ballet dancer on her points. It is found in bilateral frontal lobe disease, pseudobulbar palsy, Alzheimer's disease and normal pressure hydrocephalus.

** Jaunting gait is seen in chorea. Patient's knee may give way momentarily resulting in a fall.

*** Arthritis and muscular pain make walking stiff and slow (antalgic gait).

**** There is another Hoover's sign in internal medicine where the patients with advanced COPD show paradoxical inward movement of rib cage with each inspiration.

Different varieties of gait :

1. Neurogenic - 1, 2, 3, 4, 5 and 7
2. Myogenic - 6
3. Prosthetic - 8
4. Arthrogenic - 9
5. Psychogenic - 10

O

“Now, this is not the end. It is not even the beginning of the end. But it is, perhaps, the end of the beginning.”

— **Sir Winston Churchill (1874-1965)**

British Politician

YOUR EXAMINATION HALL

1. Be smart, gentle, polite and courteous In your gesture. Do not rush, be patient.
2. You should look confident, never overconfident; be professional in attitude.
3. Be precise. Listen the question carefully and then answer 'to the point'. Speak out clearly, slowly and concisely in a confident voice. Be optimistic and get free from any tension.
4. Do not try to be clever but be careful. Do not hesitate to revise the diagnosis.
5. Be methodical. Remember, mostly the errors are made by making cursory and incomplete physical examination than due to lack of knowledge and skill.
6. While examining a patient, you should have a lady's hand (fine and gentle touch) with an eagle's eye. Show respect to your patients as well as examiners (try to wish).
7. Nothing succeeds like thorough preparation though you may not know everything—so prepare yourself for the 'unexpected'.
8. Do not argue with your examiner. Do not be biased. Do not agree to anything you are not sure of. Be alert and focused (key of success).
9. Always try to answer in the order of frequency, i.e., from more common to less common causes, and avoid rare ones. Perform systematically.
10. Remember, the difference between success and failure is marginal.

PATH OF SUCCESS

1. Sufficient preparations with sincerity.
2. Ability to express.
3. Ability to build up good relationship between the patient and the examinee, and the examiner and the examinee.
 4. Strategic planning, creative visualisation and willpower to succeed in the face of all odds.
5. To have confidence as well as common sense, and
6. Good-luck.

INSTRUMENTS TO CARRY IN THE EXAMINATION HALL (CHECK LIST)

1. Stethoscope (preferably double-tube).
2. Hammer (preferably with flexible shaft).
3. Pencil torch (battery should be powerful).
 4. Sphygmomanometer (mercury or aneroid type); it is better to have your own instrument.
5. Pins (for testing of pain sensation and blanching reaction).
6. Cotton.
7. Measuring tape (for measurement of the abdomen in ascites, neck in thyroid enlargement, expansion of chest, head in hydrocephalus and assessment of nutrition).
8. Skin pencil.
9. Compass (for two point discrimination)—optional.
10. Tuning fork (128 or 256 cycles/sec for vibration sense, and 256 or 512 cycles/sec for cochlear function).
11. Tongue depressor.
12. Two one-rupee coin (for coin percussion in the chest, stereognosis).
13. Clinical thermometer.
14. Cotton wrapped in a broom-stick (for gag reflex).
15. Four small glass containers for taste sensation (containing solutions of sugar, common salt vinegar and quinine)—optional.
16. Cardboard or clipboard (for history writing).
17. ADMIT CARD, APRON, PEN, scale, pencil, eraser, and **CONFIDENCE**.

* ?" ^/3.hSk f°Q a/'?aCJC:reSt' (f°r neck vein examination), gloves (for per rectal examination) or eighing machine. Switch off your mobile phone while in the examination hall.

THE HISTORY SHEET

1. Prefix the patient's name by Sri/Mr/Mrs/Miss.
2. Write sequentially upto differential diagnosis. Relevant investigations are optional.
3. 'All the systems' should be written. Usually the time allotted in a 'long case' is 45 minutes.
4. In the history and clinical examination, stress the important points by underlining them.
5. Blood pressure should always be measured because it is part and parcel of general survey (mandatory—supine; optional—standing position i.e., to diagnose 'postural drop').
6. Do not write the facts which you have not enquired or examined; but remember if any test/fact is not written, it is granted that it is not done.
7. A beautiful handwriting influences the examiner. Spelling mistakes create a bad impression.
8. Try to avoid abbreviations; remember MS means mitral stenosis as well as multiple sclerosis.
9. If anaemia cyanosis or jaundice is present, mention the degree or type whichever is applicable. If any system has no abnormality, it is better to write 'within normal limit or no abnormality detected' against the subheadings of the respective system. In case of build or nutrition, the term 'average' is preferred to 'normal'. If the facies is inconclusive, write 'nothing suggestive'.
10. Write the summary meticulously. Many examiners give stress on summary. A good summary will speak for yourself.

MODEL DISTRIBUTION OF MARKS FOLLOWING RECOMMENDATION OF MCI

General Medicine : Full marks 300

Theory ; Paper I + Paper II = 60 + 60

Internal assessment : 60 (theory 30 + practical 30)

Oral (including X-ray and CT, ECG, instruments and specimens, charts, medical emergency) : 20

Clinical (bedside) : 100 (one long case 60 + one short case 30 + two spot cases 10)

* Marks distributed in the long case (60) as follows : History 15 + demonstration of signs 25 + discussion and crossing 20

** Internal assessment ; Distribution of marks (60) will be as follows ; Medicine 40 + dermatology (including STD) 10 + psychiatry 5 + chest and TB 5

*** Criteria to pass in the examination :

Aggregate 50% i.e., 150 marks

Theory and oral (including internal assessment) 50% i.e., 100 marks

Practical (bedside clinics) 50% i.e., 50 marks

N.B. ; One should get 50% in internal assessment to pass in the examination

Theory part ;

Paper I ___ CVS, respiratory system, haematology, neurology, endocrinology and rheumatology

Paper II ___ Tropical diseases, gastro-intestinal, and diseases of liver and biliary system, renal diseases, metabolic and nutritional diseases, poisoning and environmental diseases, psychiatry, dermatology including STD

**** u_{ac}h theory paper usually contains 4 short questions (4 x 10) and short notes (5 short notes x 4). Short questions are short-structured essay type and problem-oriented type. There may be overlap in

Paper I & II.

***** Acute cases (e.g., LVF, acute severe asthma) are usually not given in the examination.

Long case comprises of history writing + demonstration of signs + relevant discussion.

Short case means general survey and examination of one particular system (nothing to be written).

Spot case means spotting the diagnosis (with examiner observing) and mentioning its justification.

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Key Features of the Book

- An ideal book to recapitulate medicine before examination
- Questions and answers format with oral and practical orientation
- Model long, short and spot cases along with differential diagnosis, relevant investigations and management with stress on core topics
- Expanded coverage on clinical methods in medicine
- A concise, lucid, revised and updated ready-reckoner in medicine
- A uniquely accessible portable guide for ward and clinic



About the Author

Professor Arup Kumar Kundu has been working as an internist for nearly 25 years and is renowned for his sharp clinical acumen, analytical approach and clarity of expression towards solving different problems in internal medicine. He is vibrant and innovative in his field, and has multifaceted personality. Dr. Kundu, an astute clinician and a dedicated teacher, has been teaching medicine to students, both undergraduates and postgraduates, and has been an examiner in different Indian Universities. He is well-known all over the country for his proficiency as an illustrious teacher in medicine. Dr. Kundu, a brilliant academician and an avid medical writer, has authored three other indispensable books entitled 'Bedside Clinics in Medicine, Part II', 'MCQs in Internal Medicine' and 'Pearls in Medicine for Students'. He is a gifted orator and has been invited as speaker / faculty in different State and National conferences. He has taken part in International symposia / seminars, and is credited with more than 80 publications in peer-reviewed journals. Dr. Kundu has contributed constructive articles in National daily / weekly, interviews in AIR / Doordarshan for common people. He has written sections on online appendix of Kumar and Clark's textbook 'Clinical Medicine', both in the 6th and the 7th edition, and is an Indian member, International Advisory Panel of the 7th edition of the book. He has also contributed chapter in API textbook of Medicine, the 8th edition. Dr. Kundu, known all over as 'clinical scientist', has been conferred the fellowship of the Indian College of Physicians (FICP) and the membership of prestigious New York Academy of Sciences (USA)